Patient Resource

Anti-Glomerular Basement Membrane Disease

What is it?

Anti-glomerular basement membrane (anti-GBM) disease, also known as Goodpasture's syndrome, is one of a larger group of kidney diseases known as glomerulonephritis.

It is a rare autoimmune condition that affects the glomeruli – the tiny filters of the kidneys – and sometimes the lungs. It can lead to kidney failure if not treated promptly, and in some cases – to bleeding in the lungs.

While anti-GBM disease can affect the kidneys and the lungs, this handout will focus mainly on how it affects the kidneys.

Here's what happens in the kidneys in anti-GBM disease:

- In anti-GBM disease, the immune system mistakenly produces antibodies[1] that target a specific protein found in the kidneys and in the lungs, which can sometimes affect one or both organs.
- These antibodies are called anti-GBM antibodies. We don't know why some people develop them.
- In the kidneys, they attack a part of the filters (glomeruli) called the basement membrane, which plays a key role in filtering waste from the blood.
- This direct attack causes severe inflammation and damage in the glomeruli, leading to rapidly worsening kidney function.
- The damage in the kidneys makes them leaky, allowing blood and protein to pass into the urine.
- Because the disease can progress quickly, early diagnosis and treatment are very important to try to prevent permanent kidney damage or failure.

[1] Antibodies are protective proteins produced by your immune system. They attach to antigens (foreign substances) — such as bacteria, fungi, viruses and toxins — and remove them from your body. Source: https://my.clevelandclinic.org/health/body/22971-antibodies.

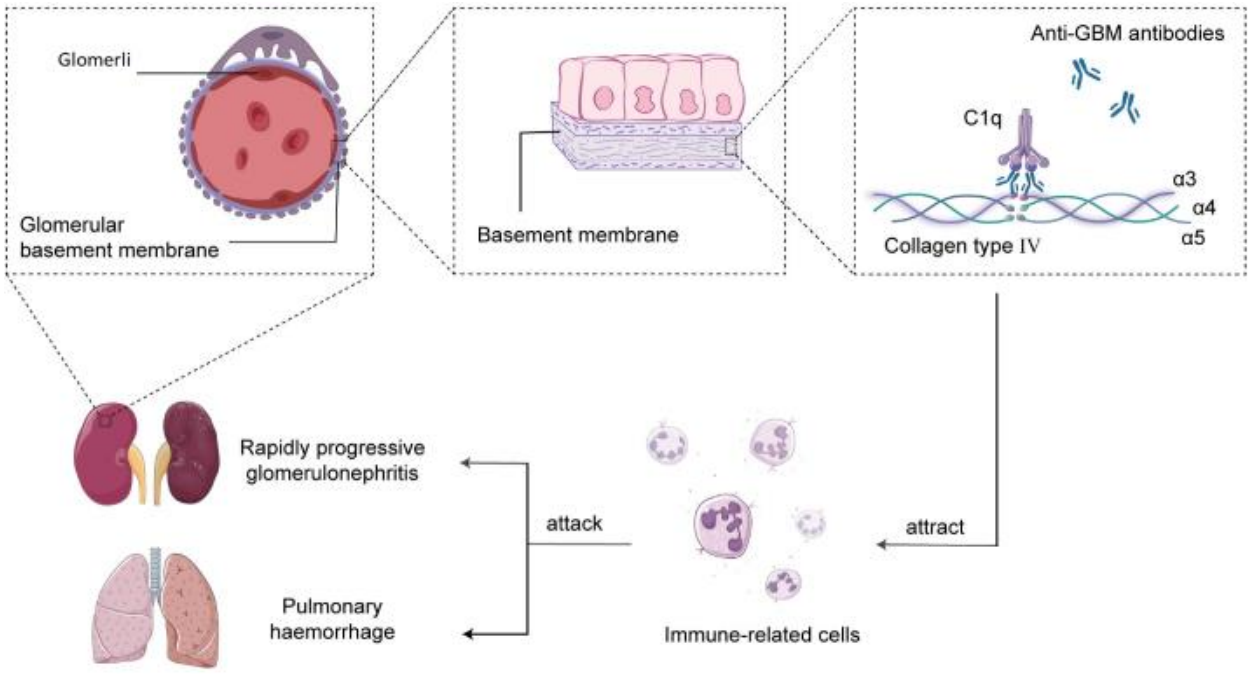

Figure source (modified): https://www.frontiersin.org/journals/immunology/articles/10.3389/fimmu.2023.1229806/full

The symptoms of anti-GBM disease may include:

- Fatigue.
- Swelling in the legs, ankles, or around the eyes.
- Foamy or bubbly urine – due to protein leaking from the kidneys.
- Blood in the urine – may look pink, red, or brown.
- Decreased urine production.
- Nausea and loss of appetite – due to worsening kidney function.
- Shortness of breath or coughing up blood – if the lungs are affected.

The complications of anti-GBM disease may include:

- Rapid loss of kidney function, which can lead to kidney failure requiring dialysis.
- High blood pressure – the kidneys help regulate blood pressure, so damage to the kidneys can cause it to rise.
- Lung hemorrhage – serious bleeding in the lungs if the disease also affects them.

What happens after you have been diagnosed with kidney disease due to anti-GBM disease?

- After your kidney biopsy is reported and a final diagnosis of anti-GBM disease 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 damage in other organs. For example, you may be referred to a respirologist.

How is anti-GBM disease treated?

The aim of treatment is to lower the production of anti-GBM antibodies and stop the immune system from causing further damage. The treatment usually begins immediately and often includes:

- Plasmapheresis (plasma exchange)[2] is a medical procedure similar to dialysis. It filters your blood to remove the anti-GBM antibodies that are attacking your kidneys and/or lungs. It is done for 2–3 weeks. If you live outside Vancouver, you may need to be transferred there for plasmapheresis, which is typically performed during a hospital admission.
- Immunosuppressive medications, which may include corticosteroids (e.g., prednisone and/or IV methylprednisolone) + one of the following:
    - Steroids.
    - Cyclophosphamide.
    - Possibly rituximab.
- The treatment duration is around 6 months, but some patients may stay longer on lower-dose medications to prevent relapse or ongoing inflammation.
- During active immunosuppressive therapy, you are at higher risk of bacterial and viral infection. If you develop a fever, please seek medical attention immediately.

General therapies:

[2] 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.

- Urine and lab tests: 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.
- Antibiotics during immunosuppressive therapy: If you are receiving immunosuppressive therapy, depending on its type, you may be started on a combination of trimethoprim and sulfamethoxazole. These antibiotics are used to reduce the risk of very serious infection that can occur in patients on medications that reduce the immune response.
- Other medications while undergoing treatment for anti-GBM disease:
    - Also, while on immunosuppression, your kidney care team may offer you medication to prevent stomach upsets.
    - While on high doses of corticosteroids, your kidney care team may recommend calcium, vitamin D, and medication to maintain bone health.
    - Medications to control swelling (diuretics) and blood pressure may be initiated depending on your symptoms.
- Medication options: Your kidney doctor and/or kidney care team will help you to learn about the medication options that would be best for you.
- Medication cost: BC Renal covers the cost of a wide range of medications used for anti-GBM treatment.

- **Important:** Patients need to check with their kidney doctor and/or kidney care team before taking any over-the-counter (OTC) medications and natural health products.
- **Important:** Patients with chronic kidney disease like anti-GBM need 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.
- **Important:** Pregnancy may impact kidney function. If you are planning a pregnancy, please consult with your kidney doctor.
- **Important:** Smoking cessation is important – it can help slow down the worsening of kidney disease. In addition, exposure to other inhaled irritants (e.g., paints) may precipitate flares of anti-GBM disease.

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Living with anti-GBM disease

- Support of kidney care team: BC kidney patients registered with BC Renal have access to a comprehensive kidney care clinic (KCC) team that includes nurses, dietitians, social workers. In most cases, patients will also have access to a pharmacist.
- Support for your wellbeing: 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.
- Ongoing follow-up: Anti-GBM disease is a serious condition that prompts immediate treatment to prevent permanent damage. Relapses are uncommon but can occur in some patients. You will need ongoing follow-up with your kidney doctor and/or kidney care team.
- Risk of kidney failure: There is a portion of anti-GBM disease patients who will end up with kidney failure, despite treatment. If your kidneys fail, your treatment options may include transplant, dialysis, and conservative care[3]. Your kidney doctor and/or kidney care team will be there to educate and support you throughout your journey.
- Participation in clinical trials: Sometimes, people living with anti-GBM disease 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.

Further information

- There may be a lot of confusing information about lupus nephritis 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/

[3] Conservative care, sometimes called conservative kidney management (CKM) or supportive kidney care, focuses on treating the symptoms of kidney failure to make you feel as well as possible. It does not treat the causes or attempt to cure kidney failure. Although conservative care does not include dialysis or transplant, it does include all of the other parts of kidney care and support from your team. For more information see this BC Renal's handout: http://www.bcrenal.ca/resource-gallery/Documents/Patient_Guide-Transitioning_to_Conservative_Care-Kidney_Care_Clinics.pdf.

- BC Renal GN web page - http://www.bcrenal.ca/health-info/kidney-care/glomerulonephritis
- Anti-GBM web page - https://www.niddk.nih.gov/health-information/kidney-disease/glomerular-disease/anti-gbm-goodpastures-disease

- 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.