

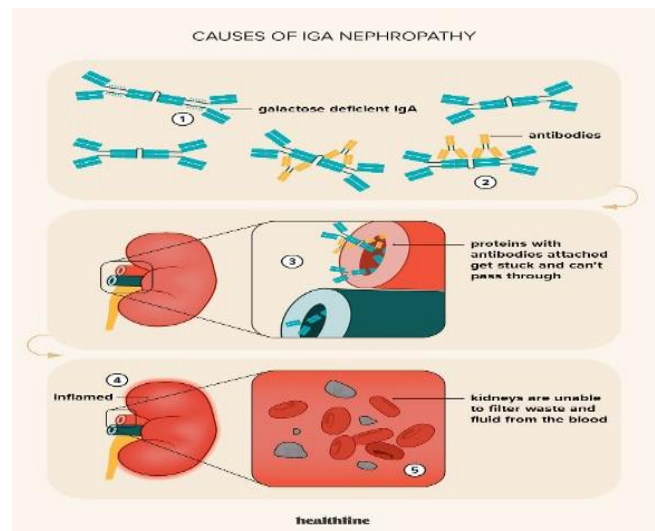
Patient Resource

IgA Nephropathy (IgAN)

What is it?

IgA nephropathy (IgAN) is the commonest autoimmune disease that affects the glomeruli – or the filters – of the kidney. IgAN is one of a larger group of kidney diseases known as glomerulonephritis. Here's what happens:

- IgA is a type of antibody¹ all people have that normally helps fight infections.
- Sometimes the immune system produces an unusual form of IgA that lacks certain sugars (galactose-deficient IgA1), making it look “foreign” to the body.
- The immune system produces more antibodies against the unusual IgA. These antibodies bind to the IgA, forming immune complexes² (clumps of proteins).
- Immune complexes travel through the bloodstream and get stuck in the tiny filters of the kidneys (glomeruli).
- The kidneys don't expect these, so their presence triggers inflammation.
- Inflammation in the kidney filters damages them over time, leading to leakage of blood and/or protein into the urine, as well as scarring in the kidneys.
- If the damage continues, the kidney filters become scarred making it harder for the kidneys to do their job. In some cases, this can lead to chronic kidney disease or even kidney failure.



Medical illustration by Maya Chastain

Figure source: <https://www.healthline.com/health/kidney-disease/igan-risk-factors-and-causes#takeaway>

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>.

² Immune complex is an antibody bound to an antigen. Immune complexes are part of a normal immune response. However, when immune complexes accumulate in the blood, they can cause autoimmune disorders. Source: <https://clinicalinfo.hiv.gov/en/glossary/immune-complex>.

Antigens are markers that tell your body that something is foreign. Your immune cells make antibodies to recognize and destroy harmful antigens. In fact, you can think of antigens as antibody generators. Source:

<https://my.clevelandclinic.org/health/diseases/24067-antigen>.

What are the symptoms of IgAN?

- Some people may have no symptoms for years, while others may notice signs early on. However, even without symptoms, the disease can cause damage.
- Here are the most common symptoms:
 - Blood in urine: sometimes it's visible (tea- or cola-colored urine), but often it's microscopic (only seen on lab tests). This can also sometimes temporarily get worse during infections
 - Frothy or foamy urine
 - High blood pressure
 - Swelling in feet, legs, hands, and eyes
 - Fatigue

What are the complications of IgAN?

- Complications may include:
 - Risk of declining kidney function, which may lead to kidney failure over time
 - Elevated cholesterol (fats) in your blood

What happens after you have been diagnosed with IgAN?

- After your kidney biopsy is reported and a final diagnosis of IgAN is made, your kidney doctor (nephrologist) and/or kidney care team may order further tests or scans and will discuss appropriate treatment with you.
- The effects of IgAN on the kidneys are variable and the path of the disease is different for each patient. For some people, IgAN may progress slowly; for others it may progress more rapidly. Your kidney care team will talk to you about the seriousness of the inflammation in your case and your treatment plan.
- In addition, there are tools your kidney doctor can use to help predict your risk of kidney function decline, and patients are encouraged to discuss this with their doctor and/or kidney care team.
- Not all IgAN is serious enough to require active treatment with medications that suppress the actions of your immune system that cause inflammation.

How is IgAN treated?

People living with IgAN may be offered a range of treatment options, from general therapies to immunosuppressive medications, with the aim of preventing further damage from the disease.

General therapies:

- Blood pressure medications (ACE inhibitors or ARBs). Even if your blood pressure is normal, they help reduce protein leakage in urine and slow down kidney damage.
- SGLT2 inhibitors. Originally for diabetes, they can help protect kidney function, even in non-diabetics, by reducing protein leakage in the urine and slowing down kidney damage.
- Statins. If cholesterol is high, statins may help reduce stroke and heart attack risk.
- Smoking cessation – quitting smoking can help slow down the worsening of kidney disease.
- 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.
- Your kidney doctor and/or kidney care team will support you to learn about the medication options that would be best for you.
- BC Renal covers the cost of a wide range of medications used for IgAN 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 IgA nephropathy 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.
- Pregnancy may impact kidney function. Individuals who are planning to have a baby should talk to their kidney doctor.

Active (immunosuppressive) therapies:

- Some people with IgAN might benefit from medications to lower the immune system response and prevent further damage to the kidney.
- During active therapy, you are at high risk of bacterial and viral infection. If you develop a fever, please seek medical attention immediately.
- Active (immunosuppressive) therapy options include:
 - Prednisone – this is the best studied treatment option.
 - Second line therapies that are not as well studied include mycophenolate mofetil and hydroxychloroquine.
 - In addition, there are various new treatment options for IgA nephropathy that have been approved in other countries but are not yet available in Canada.
- Depending on the type of immunosuppressive therapy, 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 reduce the immune response.

Living with IgAN

- 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 many patients treated for IgAN improve, the disease course is unpredictable. You will need ongoing follow-up with your kidney doctor and/or kidney care team.
- Please note that respiratory and other infections could trigger a flare in the disease, and a sign of this flare may be visible blood in the urine. Consult your doctor if you continue seeing blood in the urine after your infection resolves.
- There is a portion of IgAN patients who will end up with kidney failure despite treatment. If your kidneys fail, your treatment options may include transplant, dialysis and conservative care. Your kidney doctor and/or kidney care team will be there to educate and support you throughout your journey.
- Some people living with IgAN will be invited to participate in clinical trials for new therapies, often there are several trials available in BC. 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 IgAN and other kidney diseases on the Internet. The following websites are good sources of information for people living with IgAN:
 - 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>
 - Medication information sheets on the BC Renal GN web page - <http://www.bcrenal.ca/health-info/kidney-care/glomerulonephritis#Resources> (click on Medication information)
 - IgA Nephropathy Foundation - <https://igan.org/>
 - IgA nephropathy web page - by NephCure Kidney International - <https://nephcure.org/intro-to-rkd/types-of-rkd/iga-nephropathy-igan/>
 - IgA nephropathy web page - by GlomCon - <https://www.glomcon.org/kidney-diseases/iga-nephropathy>
 - IgA nephropathy video - by Osmosis.org - [https://www.osmosis.org/learn/IgA_nephropathy_\(NORD\)](https://www.osmosis.org/learn/IgA_nephropathy_(NORD))
- 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.