Your Guide to Understanding IgAN* Terms

Are all the medical words getting confusing? This guide explains useful terms about IgAN. It is intended to help you have an informed conversation with your doctor.

Living with IgAN? Go to KidneyHope.com

Doctor and patient portrayal.

*IgAN, immunoglobulin A nephropathy.

NOVARTIS

Table of Contents

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NOVARTIS

Kidney function and the immune system

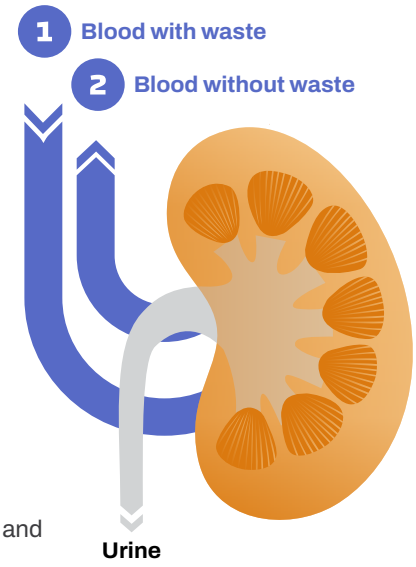
Your kidneys are vital organs; they remove waste and excess fluid from your body. At the same time, the immune system protects your body from harmful substances, germs, and unwanted changes to your cells that could make you ill.

What your kidneys do

- Filter waste and toxins from your blood
- Regulate blood pressure
- Help to make red blood cells
- Support bone health

How your kidneys work

- Each of your kidneys is made up of about a million fine filters called nephrons
- The nephrons work through a two-step process with a glomerulus and a tubule:
  - The glomerulus is made of tiny blood vessels, and it filters unneeded substances from your blood.
  - The tubule further separates the filtered fluid. It returns the useful substances back to the blood while the leftover fluid and waste becomes urine.



Your kidneys and your immune system work as a team and depend on each other to stay healthy.

By filtering toxins, the kidneys help balance vital functions of the immune system. At the same time, your immune system protects your kidneys from foreign elements that could disrupt their function and can play an important role in chronic kidney diseases, like IgAN.

What is IgAN?

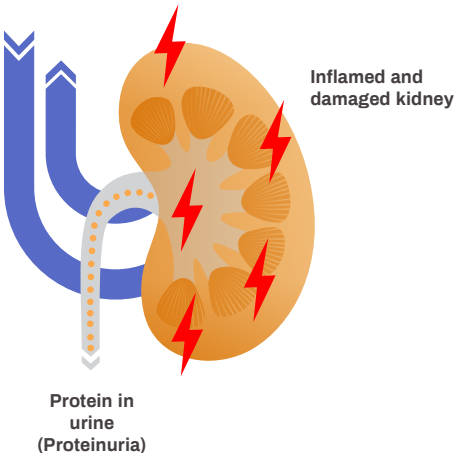
Immunoglobulin A nephropathy (IgAN), also called Berger's disease, is a progressive, long-lasting autoimmune disease that attacks abnormally high levels of an IgA protein. This can result in kidney damage and function decline.

The journey with IgAN looks different for every patient.

How IgAN impacts kidney health

When you have IgAN, your body produces abnormally high levels of an IgA protein. Your immune system then attacks these proteins, causing them to form clusters that accumulate in the kidneys.

This accumulation can result in inflammation and damage that can affect how well your kidneys work.



How IgAN can lead to kidney function decline

There are several ways IgAN can damage your kidneys:

- One way is through the **endothelin A (ET_A) pathway**, which is involved in regulating multiple functions in the kidney like blood pressure and balancing concentrations of salts and fluid. In IgAN, the ET_A pathway is turned on, and it can lead to kidney damage and scarring through various processes. As a result, protein can leak into the urine, leading to proteinuria.
- Another way is through a key part of the immune system called the **complement system**. When you have IgAN, your complement system can become overactive in your kidneys. This can result in inflammation and injury.

The risk of proteinuria in IgAN

When your kidneys are damaged or not working properly, protein can leak into your urine. This is known as proteinuria.

Proteinuria can be a sign that your kidneys aren't working the way they should and can mean kidney function decline. In people with IgAN, managing proteinuria is an important goal. That's why it's important for you and your doctor to monitor your protein levels and how they change over time.

Everyone with IgAN is different, and a variety of factors can impact IgAN progression. Retrospective studies show that people with IgAN who have ongoing proteinuria may be at risk of their disease worsening. Additionally, those who have higher levels of proteinuria are more likely to experience a more rapid loss of kidney function. **According to global expert guidelines on IgAN, if your proteinuria is more than 0.75-1 g/day, you may be at high risk for your IgAN to get worse.**

A retrospective study* reported the percentage of people with IgAN progressing to kidney failure within 10 years of diagnosis, based on different levels of ongoing proteinuria.

Ongoing proteinuria	Estimated % of people progressing to kidney failure within 10 years of diagnosis
Less than 0.5 g/day	22%
0.5 to less than 1 g/day	31%
1.0 to less than 2 g/day	60%
2 g/day or more	85%

*A retrospective study means the data were collected in the past and not intended to be analyzed for research purposes. There were additional limitations to this UK-based study, including a lack of data about patients' medications and blood pressure.

Talk to your doctor about your proteinuria goals.

Glossary

Antibodies
Proteins produced by your immune system to protect your body from unwanted substances.

Autoantibodies
These antibodies mistakenly target your own body. This could trigger damage to different parts of the body, including the kidneys.

Autoimmune disease
When your body's natural defense system mistakenly attacks your own body.

The complement system
This is a part of the immune system that helps remove unwanted substances from the body. In IgAN, activation of the complement system can worsen inflammation and damage in the kidneys.

ET_A pathway
The ET_A pathway is activated by the ET_A receptor, a protein that is thought to play a role in the progression of IgAN. In IgAN, activation of the ET_A pathway can lead to a number of effects, such as damage and proteinuria.

Immunodeficiency
When your immune system can't adequately protect your body from infection.

Immunoglobulin A (IgA)
IgA is an antibody. In IgAN, your immune system produces more of a specific type of IgA protein than normal. This may trigger the body to make autoantibodies, resulting in the autoimmune disease.

Proteinuria
Protein in the urine, which is a signal that kidneys aren't working properly.

Renal
Anything related to the kidneys.

Still exploring the terms?
Find ways to discuss your condition.
Find the support you need at
KidneyHope.com

The stages of kidney disease

Become familiar with the kidney disease stages to better understand your condition.

How kidney disease may progress

The 5 stages of kidney disease are determined by kidney function, or how well your kidneys are working. This is measured by a test called glomerular filtration rate (GFR).

Stage	1	2	3	4	5
Kidney function	**Slight kidney damage** with normal kidney function	**Mild loss** of kidney function	**Mild, moderate, or possibly severe** loss of kidney function	**Moderate to severe** kidney damage, loss of function	**Advanced** kidney disease, or end-stage disease
% of normal kidney function based on GFR	**90% or higher**	**60% - 89%**	**30% - 59%**	**15% - 29%**	**Less than 15%**
Symptoms	Usually none		Possible changes in urine and swelling of hands and feet	All symptoms of stage 3, plus possible shortness of breath	Includes symptoms from earlier stages, plus difficulty breathing or sleeping, feeling very weak, chest pain, difficulty concentrating, nausea, or vomiting
Management and treatment	**Supportive care** with management of blood pressure and lifestyle changes are recommended. In certain patients, management of blood sugar and cholesterol levels may also be considered to help keep kidney disease from worsening.		**Regular visits** with your nephrologist can help you manage kidney disease. Your doctor can take steps for addressing kidney damage and possible treatments for advanced chronic kidney disease, including bone disease or anemia.		**Dialysis and transplantation** may be considered.

GFR, glomerular filtration rate.

Glossary

Asymptomatic

When you show no symptoms of disease. Without showing symptoms, it can be difficult to get a diagnosis. For example, if you've been diagnosed with IgAN, your disease may be progressing even though you may not feel the symptoms.

CKD

Chronic kidney disease. This is the gradual, ongoing loss of kidney function.

Dialysis

A procedure during which a machine outside the body is used to remove waste products and excess fluid from the blood when the kidneys stop working properly. Each treatment usually takes about 3 to 5 hours, with treatment repeated 3 times per week. Patients may experience a drop in blood pressure. If this happens, they may feel sick to the stomach, or have a headache or cramps. The two main types are (1) hemodialysis, in which the blood is filtered by a machine; (2) peritoneal dialysis, in which a fluid is put into the abdomen so that wastes can dissolve into it. Then the fluid is replaced or removed.

Edema

Swelling caused by excess fluid trapped in your body's tissues. Swelling in the body often increases when a person has heart or kidney disease that is getting worse.

ESKD/ESRD

End-stage kidney disease, also called end-stage renal disease. This is the last stage of chronic kidney disease, when the kidneys can no longer support the body's needs, and dialysis treatment or a kidney transplant is required.

Fatigue

A feeling of being tired and weak, often caused by a severe decrease in kidney function.

Renal hypertension

High blood pressure caused by the narrowing of the arteries that carry blood to the kidneys. When the kidneys don't get enough blood, they react by making a hormone that makes the blood pressure rise.

Kidney transplant

A serious surgery for implanting a kidney from a living or deceased donor into a person with end-stage kidney disease.

Discover more about IgAN symptoms and management.
Find the support you need at
KidneyHope.com

Testing kidney function

Learn about tests and what information they can provide about IgAN.

Lab test	Why is it relevant?	What is the procedure?	What is the goal?
Urine protein-to-creatinine ratio (UPCR) A measurement of protein in urine, or proteinuria.	Ongoing proteinuria in a person with IgAN may be a sign of progressive kidney damage that can lead to kidney failure.	Tested using a urine sample and measured in grams per day (g/day) or grams per gram (g/g). 0.88 g/g is approximately 1 g/day.	Testing for proteinuria gives a reliable indication of disease progression. If you continue to have protein in your urine, talk with your doctor about your concerns.
Hematuria Checking for the presence of blood in the urine.	You may get blood in your urine because of kidney inflammation.	Tested using a urine sample.	If test results continually show blood in your urine, you may need more tests. Talk to your doctor about what this means, and if it affects your care plan.
Blood pressure (BP) The amount of force your blood pushes against the walls of your blood vessels.	High blood pressure can damage the kidneys and reduce kidney function.	Measured using a cuff.	According to professional guidelines, your target systolic blood pressure should be 120 mmHg.
Estimated glomerular filtration rate (eGFR) A calculation used to assess kidney function, or how well your kidneys work.	Assessing eGFR along with proteinuria can help determine the stage of kidney disease. Rapid eGFR decline can be associated with faster IgAN worsening.	Tested using a blood sample.	The goal is to maintain kidney function. This is indicated by a stable or improving eGFR.
Biopsy Taking one or more little pieces of the kidney to examine under a microscope.	A biopsy is the only definitive way to diagnose IgAN, but it may not be an option for everyone.	A sample of kidney tissue is taken either using a needle guided by ultrasound or directly during surgery.	Results establish IgAN diagnosis. A biopsy can also help your doctor determine how serious your IgAN is, or how quickly your IgAN may progress.

Get more informed about your condition

Knowing these frequently used terms may help you better navigate your treatment journey.

Medications

ACEi/ARB

Angiotensin-converting enzyme inhibitors (ACEi) and angiotensin receptor blockers (ARB) are medications used to help reduce blood pressure and proteinuria, slow progression of kidney disease, and improve outcomes in patients who have heart failure, type 2 diabetes, or a history of heart attacks.

Corticosteroids

Medications used to treat a variety of inflammatory diseases.

SGLT2 blockers

Sodium-glucose cotransporter-2 blockers are medications given to certain patients to lower their blood sugar.

The IgAN treatment landscape is rapidly changing. Be sure to ask your doctor about all the treatment options available to you.

Nutrition guidelines

Fish oil, omega-3 fatty acids

Supplements used to help prevent kidney inflammation and slow the progression of kidney disease. Be sure to consult your doctor before taking fish oil, omega-3 fatty acids, or any other supplements.

Kidney-friendly diet

Your doctor may prescribe a food plan to help you manage your kidney disease and slow damage to your kidneys. This diet will help to keep certain minerals from building up. This is important because with IgAN your kidneys do not work as well to remove waste products from your body.

Living with IgAN?
Get support at **KidneyHope.com**

Glossary

Health care team

Dietitian/Nutritionist
A medical professional who can suggest dietary modifications to help your body better cope with IgAN.

Nephrologist
A medical doctor who diagnoses and treats kidney diseases.

Pathologist
A medical doctor who reviews tissue from the body under special microscopes to diagnose diseases.

Primary care physician
A general medical doctor who may refer you to a specialist such as a nephrologist.

Health insurance

Co-insurance
The percentage of covered medical costs you will pay after you've reached your deductible.

Co-pay
A fixed amount (for example, $20) you pay for a covered health care treatment or service after you've met your deductible.

Deductible (Ded)
The total amount you pay for covered health care treatment or services before your insurance plan starts to pay.

Explanation of benefits (EOB)
The insurance company's written explanation regarding a claim, showing what they paid and what the patient must pay.

Glossary (continued)

In-network (INN)

When a doctor, hospital, or other provider accepts your health insurance plan, they are in-network.

Maximum out-of-pocket (MOOP)

The most you must pay for covered services in a plan year.

Prior authorization (PA)

A requirement by health plans for patients to obtain approval for a health care service or medication before it is provided.

Find other helpful resources, like a **Symptom Tracker** and **Productive Appointment Guide** on the resources page of **KidneyHope.com**

Talk to your doctor about your IgAN and how you can manage it.

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