

RARE KIDNEY DISEASE 101

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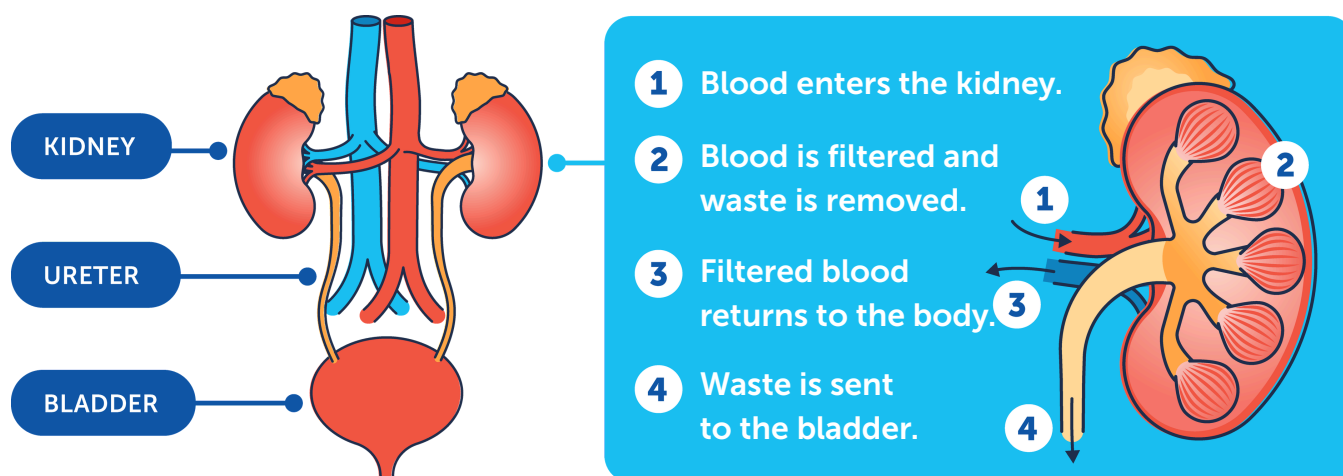
About This Guide

This guide was created to help patients and families navigate a rare kidney disease diagnosis with confidence. Whether you were recently diagnosed with C3 glomerulopathy (C3G), immune complex-mediated membranoproliferative glomerulonephritis (IC-MPGN), or another rare kidney disease, this guide will help you understand your condition, know what to expect, and feel more prepared for conversations with your care team.

SECTION 1

Understanding Your Kidneys and Your Diagnosis

Your kidneys do far more than filter waste. Every day, they regulate blood pressure, balance fluids and electrolytes, produce hormones that support red blood cell production, and keep your body's chemistry in check. When the kidneys are damaged — especially by a rare disease — these functions can all be affected.



What Makes a Kidney Disease 'Rare'?

The U.S. Food and Drug Administration (FDA) defines a rare disease as one affecting fewer than 200,000 people in the United States. Some examples of rare kidney diseases include:

- C3 glomerulopathy (C3G)
- Immune complex-mediated membranoproliferative glomerulonephritis (IC-MPGN)
- Immunoglobulin A nephropathy (IgAN)
- Focal segmental glomerulosclerosis (FSGS)
- Alport syndrome

Despite their differences, these conditions share a common thread. They are frequently underdiagnosed or misdiagnosed, often because their early symptoms overlap with more common conditions.

Why Diagnosis Takes Time

Many people with rare kidney diseases see multiple specialists before getting a definitive answer. Lab results may be abnormal for months or years before a specific diagnosis is made. If that's been your experience, you're not alone. Getting to the right diagnosis matters because treatment for a rare kidney disease may be different from standard chronic kidney disease (CKD) management. CKD is a broad term for kidneys that have lost function over time, regardless of the underlying cause — it describes what's happening, not why.

SECTION 2

Your Diagnosis Explained

Understanding your diagnosis means understanding a few key tests and what they reveal.

How Kidney Function Is Measured

Your care team uses several markers to assess kidney health:

- Estimated glomerular filtration rate (eGFR) — This estimates how well your kidneys are filtering blood. A normal eGFR depends on your age; lower numbers indicate reduced function.
- Creatinine — This waste product builds up in the blood when the kidneys aren't filtering well.
- Protein and blood in urine — Both signal that the kidney's filtering system is damaged, even when you can't see or feel it. This may be the first sign of kidney disease.

Understanding Your Stage

Kidney disease is staged 1 through 5 based on eGFR and signs of kidney damage:

Stage 1-2

Normal or mildly reduced function with early signs of damage

Stage 3

Moderate reduction in kidney function

Stage 4

Severe reduction, preparation for kidney replacement therapy may begin

Stage 5

Kidney failure

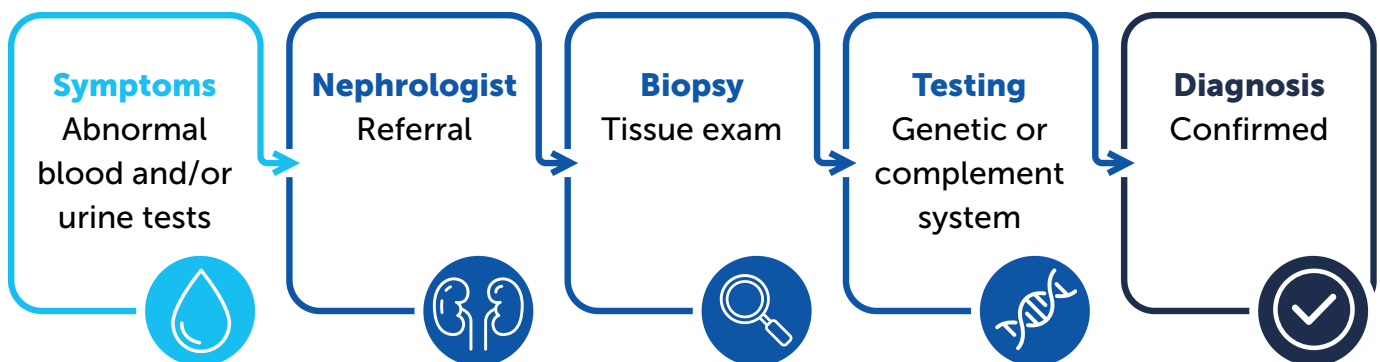
Some people maintain a relatively normal eGFR even while active inflammation is occurring. Staging gives your care team a baseline, but it's one piece of a larger picture.

What a Biopsy Reveals

A kidney biopsy may be the most important step in diagnosing a rare kidney disease. It involves removing a small sample of kidney tissue to examine under a microscope. For conditions like C3G and IC-MPGN, the biopsy can identify specific patterns of inflammation that distinguish these diseases from other kidney conditions — and from each other. Without a biopsy, many rare kidney diseases are impossible to diagnose accurately.

Why the Diagnostic Journey Takes Time

Rare kidney diseases are often initially mistaken for common CKD. Genetic testing, complement system testing, and a specialist review of biopsy results may all be needed before a final diagnosis is confirmed. Here's what that journey typically looks like:



Months to years: It is common to wait months, or longer, for a confirmed diagnosis. Each step requires specialist review and can take time to arrange.

SECTION 3

Understanding Your Symptoms — CKD, Rare Kidney Disease, and What's Causing What

Many patients with rare kidney diseases are also diagnosed with CKD — or were told they had CKD before getting a more specific diagnosis. Understanding the relationship between the two helps make sense of your diagnosis.

CKD vs. Rare Kidney Disease: What's the Difference?

CKD describes the degree of kidney damage over time — how well the kidneys are working, but not why they're damaged.

A rare kidney disease like C3G or IC-MPGN can be the underlying cause. The rare disease can lead to CKD as kidney function declines, which is why both diagnoses sometimes appear together.

This distinction matters for treatment. Standard CKD management focuses on slowing damage through blood pressure control, diet, and supportive medications. That's necessary, but not sufficient if you have an active rare kidney disease driving the damage. Treating the underlying cause may require targeted therapies specific to your disease. If you received a CKD diagnosis before your rare kidney disease was identified, it's worth revisiting your treatment plan with a nephrologist who specializes in rare kidney diseases.

Why Symptoms Overlap

Because rare kidney diseases and CKD so often coexist, their symptoms frequently overlap. Some symptoms are more specific to rare kidney diseases; others are shared; and some come from co-occurring conditions entirely.

SECTION 4

Your Care Team

Managing a rare kidney disease rarely involves just one doctor. Here's who you may work with and what each person does.

Your Core Team Members



Nephrologist

Your lead specialist who manages kidney function, interprets lab results, and oversees your treatment plan



Urologist

Addresses structural or urinary tract issues that may accompany kidney disease



Dietitian or nutritionist

Helps you plan a kidney-friendly diet, including what to limit and why



Pharmacist

Reviews your medications for kidney safety, since many common drugs are processed by the kidneys and can cause harm at normal doses



Social worker

Helps navigate insurance, disability paperwork, and transplant logistics



Mental health provider

Supports you through the anxiety, depression, and adjustment challenges that often come with a chronic illness diagnosis



Primary care physician

Coordinates your overall care and keeps other providers informed



Advanced practice provider

A nurse practitioner or physician associate who may help the nephrologist with your care by seeing you in the office or answering your questions



Nurse

Helps support the nephrologist and advanced practice provider

Coordinating Your Care

With multiple providers involved, communication gaps are common. A few things that help:

- Keep a single running document with your current medications, recent lab values, and upcoming appointments. Bring it to every visit.
- Ask each provider to copy others on notes or letters.
- Don't assume one specialist knows what another has recommended. You may need to be the link between them.
- If possible, keep all of your specialists in one hospital system so that communication is easier.

SECTION 5

Questions To Ask Your Doctor

You may not get through all of these in one visit. Choose the ones that matter most to you right now, write them down, and bring them to your next appointment.

At Diagnosis

- What specific type of rare kidney disease do I have?
- How confident are you in this diagnosis?
- Do I need a biopsy, and what will it tell us?
- Are there genetic factors I or my family should know about?

About Your Treatment Plan

- What are my treatment options, and what are the trade-offs?
- Are there clinical trials I might qualify for?
- How will we know if treatment is working?
- Which of my current medications could be harmful to my kidneys?

About Monitoring and Progression

- How often should I be seen and tested?
- What symptoms or lab changes should prompt me to call you?
- What does disease progression look like in my specific condition?
- When should we start talking about dialysis or transplant?

About Daily Life

- Are there foods, supplements, or activities I should avoid?
- If another provider recommends a medication, how do I check to see if it's safe for my kidneys?
- Can I still exercise, travel, and work?
- What should I tell other providers I see?

About Communication

- What is the best way to communicate with you?
- How will you let me know about test results?
- How should my other providers communicate with you if they have questions?

SECTION 6

Lifestyle and Self-Management

Diet

Depending on your kidney disease and your CKD stage, you may have to limit certain parts of your diet:

Protein

Produces waste the kidneys must filter (especially animal protein)

Sodium

Drives fluid retention and raises blood pressure

Potassium

Can build up in the blood (found in bananas, potatoes, tomatoes, and many other foods)

Supplements are a particular concern — many over-the-counter products contain ingredients that stress already-compromised kidneys. Always check with your care team before adding anything new.

Exercise

Staying active is important with kidney disease. Regular moderate exercise can help manage blood pressure, weight, and fatigue. What's appropriate depends on your current kidney function and symptoms — ask your nephrologist what level of activity is safe for your specific situation.

Sleep and Stress

Sleep disturbances are common in kidney disease. Conditions like restless legs syndrome and sleep apnea occur more frequently in people with CKD, and poor sleep worsens fatigue, blood pressure, and mental health. Raise sleep issues with your care team rather than accepting them as inevitable.

For stress, evidence-based approaches include:



**Mindfulness and
meditation practices**



**Cognitive behavioral
therapy (CBT)**



**Peer support through
patient communities**

Monitoring Yourself at Home

- Track your blood pressure regularly and bring the log to appointments.
- Watch for signs of fluid retention: swelling in your legs, sudden weight gain, or increased shortness of breath.
- Know your red-flag symptoms — ask your nephrologist for a specific list for your condition.
- Know when to call your care team and when to go to the emergency room.

Glossary

Antibody

A protein that is made by the immune system to identify and neutralize threats like bacteria and viruses.

Biopsy

This procedure, in which a small sample of tissue is removed and examined under a microscope, helps identify disease.

C3 glomerulopathy (C3G)

This rare kidney disease is caused by abnormal regulation of a part of the immune system called the complement system.

Chronic kidney disease (CKD)

This is a broad term for kidneys that have lost function over time. CKD describes the degree of kidney damage, not its underlying cause. A rare kidney disease can cause CKD as it progresses.

Complement system

This part of the immune system helps fight infection. Abnormal complement activity plays an important role in C3G and may also be involved in IC-MPGN.

Creatinine

This is a waste product filtered by the kidneys. Rising creatinine levels in the blood signal declining kidney function.

Edema

This swelling is caused by excess fluid trapped in body tissues. In kidney disease, it most commonly appears in the legs, feet, and ankles.

Estimated glomerular filtration rate (eGFR)

This is a measure of how well the kidneys are filtering blood. A normal eGFR is dependent on your age, but for most young adults, 90 or higher is typical. Lower numbers may indicate reduced kidney function. eGFR is used to help stage kidney disease from 1 (mild) to 5 (kidney failure).

Focal segmental glomerulosclerosis (FSGS)

In this rare kidney disease, scar tissue forms in the glomeruli, the tiny filtering units of the kidney.

Glomeruli

These are tiny filtering units inside the kidneys where blood is cleaned and waste is separated out. Many rare kidney diseases damage the glomeruli directly.

Immune complex-mediated membranoproliferative glomerulonephritis (IC-MPGN)

This rare kidney disease is caused by an active immune system that leads to inflammation in the glomeruli and kidney damage.

Immunoglobulin A nephropathy (IgAN)

In this rare kidney disease, also called Berger's disease, IgA antibodies accumulate in the kidneys and cause inflammation.

Nephrologist

A doctor who specializes in kidney disease, your nephrologist is typically the lead specialist in managing a rare kidney disease.

Proteinuria

Proteinuria, or excess protein in the urine, is a common sign that the kidney's filtering system is damaged.

Renal

This term means "relating to the kidneys." "Renal function" means kidney function and "renal disease" means kidney disease.