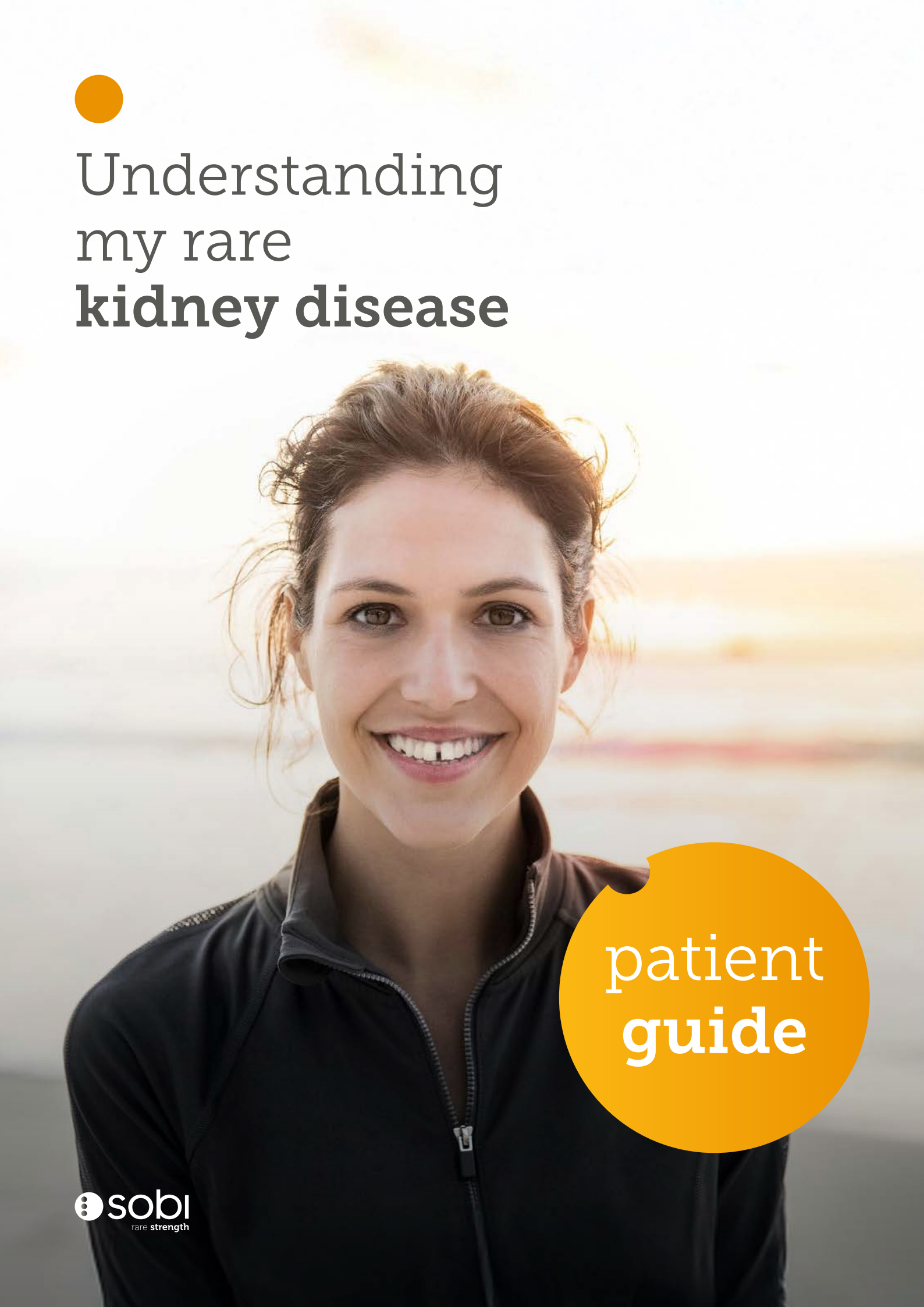
Understanding my rare **kidney disease**



patient **guide**

sobi
rare strength

Welcome to your guide

Living with a rare kidney disease such as C3 glomerulopathy (C3G) or primary (idiopathic) immune complex-mediated membranoproliferative glomerulonephritis (IC-MPGN) can feel overwhelming. This guide is designed to help you better understand your diagnosis and empower you to confidently discuss your needs and care plan with your healthcare team.

About 1 to 3 new cases of C3G are diagnosed each year for every one million people worldwide, and primary IC-MPGN may be even rarer.[2]

What are C3G and primary IC-MPGN?

C3G and primary IC-MPGN are two different ultra-rare kidney diseases that share a common issue: an overactive C3 protein in the immune system.

Normally, C3 helps protect the body from infections, but when it becomes overactive, it can mistakenly damage the kidneys. This happens by affecting the glomeruli—the tiny filters in the kidneys responsible for removing waste and excess fluid.

Despite being distinct conditions, C3G and primary IC-MPGN have overlapping symptoms, including the triad of:[1]

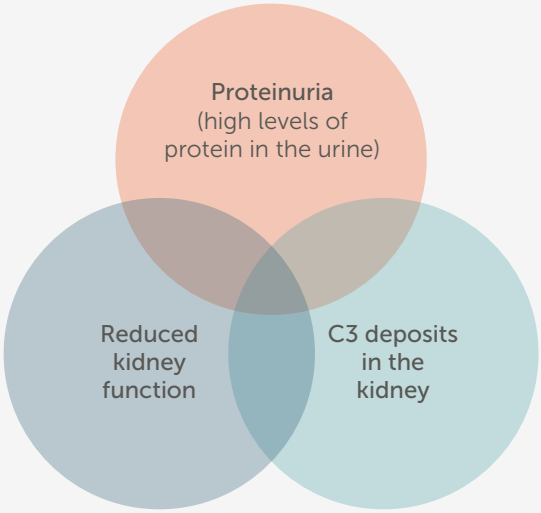
— Proteinuria (high levels of protein in the urine)

— Reduced kidney function

— C3 deposits in the kidney

While C3G and primary IC-MPGN are similar, they are caused by different underlying processes in the body.

Finding these differences in a biopsy helps your doctor provide an accurate diagnosis so that together you can choose the best care plan for you.



C3G[1,3]

C3G occurs when an abnormal amount of the C3 protein is found in the kidneys without significant antibody involvement.

IC-MPGN[1,4]

IC-MPGN occurs when both C3 and antibodies build up in the kidneys.

Who gets C3G and primary IC-MPGN?

If you're wondering, "Why did this happen to me?", you're not alone.

These conditions can affect anyone, but they are most commonly diagnosed in childhood or early adulthood. They affect males and females equally, and in most cases, there is no clear reason why they develop. However, some people may have a higher risk if they have:[1]

— A family history of kidney disease.

— Certain genetic factors that affect how their immune system and kidneys work.

While certain factors may contribute to disease development, these are not things you could have controlled or prevented.

What are the signs and symptoms of C3G and primary IC-MPGN?

Both C3G and primary IC-MPGN disrupt the kidney's ability to filter blood effectively, leading to similar signs and symptoms:[1]

Proteinuria is a key symptom, and easy to test for and monitor progression.

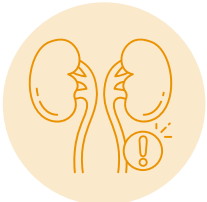
Proteinuria
Excess protein in the urine, often causing foamy or frothy urine.

Swelling (oedema)
Fluid retention, particularly in the legs, ankles, hands, and around the eyes.

Hypertension (high blood pressure)
A common symptom that can worsen kidney damage over time.

Haematuria (blood in the urine)
Urine may appear pink, red, or brown, or microscopic blood may be detected during testing.

Fatigue and weakness
Caused by the build-up of waste products in the blood and sometimes worsened by anaemia.



Decreased kidney function
Often identified through blood tests, such as elevated creatinine levels, and can progress to kidney failure in severe cases.

Symptoms can vary depending on the stage of the disease and any underlying conditions that might be contributing to the kidney damage.

How are C3G and primary IC-MPGN diagnosed?

People showing signs and symptoms of kidney dysfunction may be asked to go through a range of diagnostic procedures:[1]

This may feel like a lot of tests, but they are essential for getting a clear and accurate diagnosis. It's understandable that some people may feel hesitant about a kidney biopsy, but it is the only reliable way to confirm C3G or primary IC-MPGN. Having this clarity ensures you receive the right care and a treatment plan tailored to your needs.

Medical history and **physical examination** to assess symptoms, family history, and underlying conditions.

Urine and blood tests to test for proteinuria and haematuria.

Kidney biopsy to assess structural changes in the kidney and complement or immune complex deposits.

Genetic and autoantibody tests to identify abnormalities in the complement system that can contribute to kidney damage.

For primary IC-MPGN, **additional tests** are conducted to rule out secondary causes and underlying conditions.

Are C3G and primary IC-MPGN progressive?

Yes, both C3G and primary IC-MPGN are chronic progressive diseases and, if left untreated, may result in irreversible damage.

The good news is that early diagnosis allows for timely treatment, which can allow for a timely and individualised care plan to be put in place.[5]

50%

Up to–50% of people with C3G and primary IC-MPGN experience kidney failure within 10 years of diagnosis. This requires dialysis or kidney transplantation.[1]

What self-management steps can I take?

Here are some suggestions to help you to feel more in control of your condition:[6]

— Speak to your healthcare provider about any dietary adjustments you can make and what physical activity might be best for you.

— Take care of your emotional well-being by talking to trusted family or friends. If you feel you need more emotional support or want to learn new coping skills, explore your options for speaking to a trained counsellor.

— Schedule regular follow-ups with your nephrologist, ensuring you discuss any new signs, symptoms or concerns you might have. That's what they are there for!

Where can I learn more about my condition?

If you would like to learn more about C3G and primary IC-MPGN, there are other resources available for you:

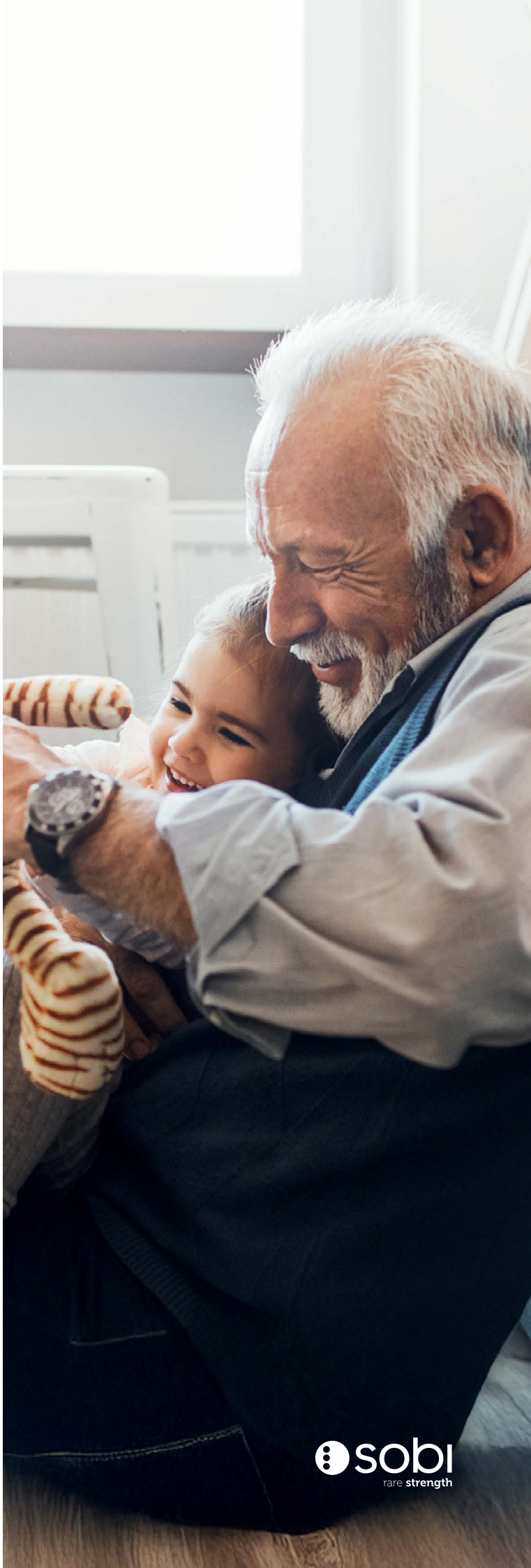
World Kidney Day
worldkidneyday.org

My C3G-ICMPGN
myc3g-icmpgn.com

Nephcure
nephcure.org

Compcure
compcure.org

It's equally important to communicate openly with your healthcare team—ask questions, share your concerns, and discuss your thoughts to ensure the care you receive is meeting your needs.



Swedish Orphan Biovitrum AB (publ) SE-112 76 Stockholm, Sweden, +46(0)8 697 20 00
Date of preparation: April 2025

References
1. Bomback, A.S., Charu, V., and Fakhouri, F. (2025) Challenges in the Diagnosis and Management of Immune Complex-Mediated Membranoproliferative Glomerulonephritis and Complement 3 Glomerulopathy. Kidney International Reports, 10(1), pp. 17-28.
2. Orphanet (2024). C3 glomerulopathy. Available at: www.orpha.net/en/disease/detail/329918
3. Caravaca-Fontán, F., Lucientes, L., Cavero, T., and Praga, M. (2020). Update on C3 Glomerulopathy: A Complement-Mediated Disease, Nephron, 144(6), pp. 272–280.
4. Noris, M., Donadelli, R. & Remuzzi, G. (2019). Autoimmune abnormalities of the alternative complement pathway in membranoproliferative glomerulonephritis and C3 glomerulopathy. Pediatr Nephrol, 34, pp. 1311–1323 (2019).
5. Caravaca-Fontán, F., Toledo-Rojas, R., Huerta, A. et al. (2025). Comparative analysis of proteinuria and longitudinal outcomes in immune complex membranoproliferative glomerulonephritis and C3 glomerulopathy. Kidney International Reports. In press: www.sciencedirect.com/science/article/pii/S246802492500049X
6. ERKNet - C3G and IC-MPGN (2025). Available at: www.erknet.org/patients/your-kidney-disease/c3g-ic-mpgn/disease-informationwww.erknet.org/patients/your-kidney-disease/c3g-ic-mpgn/disease-information