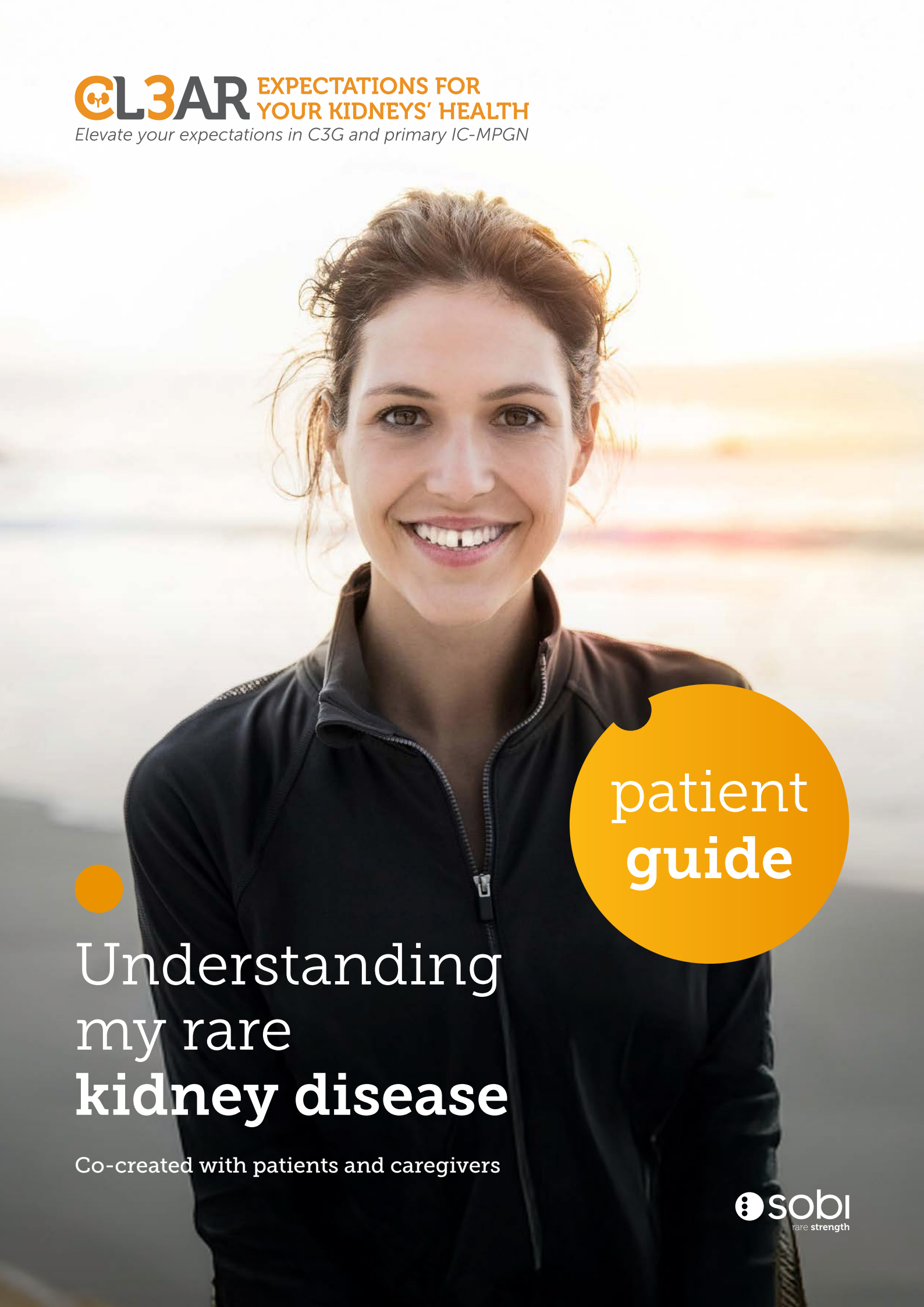
CL3AR EXPECTATIONS FOR YOUR KIDNEYS' HEALTH

Elevate your expectations in C3G and primary IC-MPGN



patient **guide**

Understanding my rare **kidney disease**

Co-created with patients and caregivers

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]

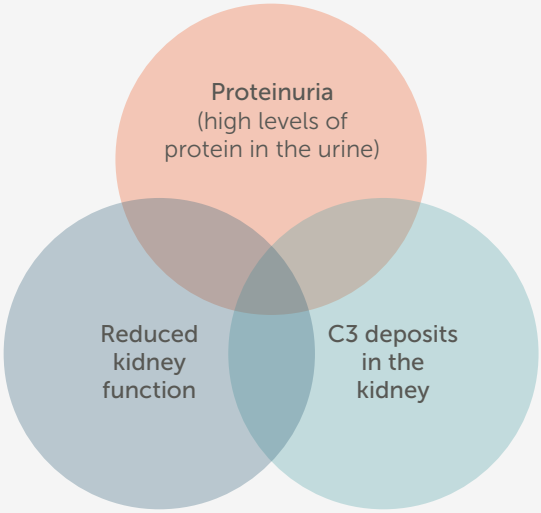
— Proteinuria (high levels of protein in the urine)

— 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.[5]

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. Some patients may experience anxiety about kidney biopsy, which is a common response to invasive diagnostic procedures, but it is the only reliable way to confirm C3G or primary IC-MPGN.[6] 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 complement 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.[7]

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:[8]

— 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 family or friends. You can also contact your local patient organisation. If you feel you need more emotional support or want to learn new coping skills, explore your options for speaking to a trained counsellor or a patient support group.

— 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!

Making the most of your healthcare appointments

It's a good idea to prepare for appointments with your nephrologist. You can ask the questions that are relevant to you, to help you understand your condition better.

Your nephrologist can help you manage your symptoms and, ultimately, help improve your quality of care and life.

Tips to help you prepare for your next appointment

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1. Prioritise questions you may want to ask your healthcare team. Get yourself a dedicated notepad to write down questions as they arise, so you don't forget them and can take them with you to your appointment.
2. Be open about your symptoms.
3. Share any changes you notice in your physical wellbeing, such as weight loss, changes in urination, nausea, headaches or dizziness, or other conditions that have changed since your last visit
4. Share any changes you notice in your emotional wellbeing, such as feeling more anxiety, sadness, stress or frustration, or improvements in your state of mind.
5. If in doubt about lifestyle changes you can make, ask your nephrologist for advice.
6. Ask about your test results and what they mean.
7. Inform your nephrologist of any changes that may impact your care plan (like forgetting to take your medication, experiencing an infection/illness or changes in diet).
8. Ask for the support you need.
9. If you are currently in dialysis, be open with your nephrologist about your life goals and expectations. Feel free to express your needs and priorities for the future and ask your nephrologist for care and life approaches that fulfil your needs. This is called shared decision making.
10. If you live with a transplanted kidney and have concerns or doubts about specific care approaches, be open with your nephrologist about what you expect for your future in terms of kidney health and possible options.

Questions you may want to ask your nephrologist

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Questions that may be helpful at initial consultations

Can you explain my disease and what is happening in my body?

What are my care options? How can we decide, together, on the optimal care plan for me?

What outcomes are you expecting from my care plan and why?

How much experience do you have treating my rare condition?

Know that it's okay to ask questions about your care plan options so that you can be fully involved in the decision-making process. This is called 'shared decision making' (SDM).

Questions about monitoring progress

What values are you monitoring while taking care of me? Which of these values are the most important and why?

What are good values to have a satisfactory chance of preserving my kidney function?

Can you talk me through my recent test results and what this means for me?

Based on these results, is there anything I need to do?

Remember, your healthcare team are there to support and understand your needs. Be sure to highlight what matters to you most during your appointments.

Questions about everyday living

Should I be making any changes to what I eat or drink?

Are there any types of exercise you would recommend?

Is there any habit I should start/continue/stop to improve my care and life?

Questions to reflect on:

Which signs or symptoms have I noticed since my last appointment? Examples include proteinuria (protein in urine), swelling (particularly — but not limited to — in the legs, ankles, and around the eyes), blood in urine, fatigue and general weakness, or other)

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Which symptoms have impacted my life the most (e.g. affected my relationships with friends/family/colleagues, led to days missed from work or school)?

How is my rare kidney disease impacting me emotionally and mentally?

Is anything specifically worrying me about my condition?

Is there anything that my disease is stopping me from doing?

What are my goals and expectations for the future?

Am I happy with my current care plan? Are there care options I may not have tried yet?

Is there any care approach that could help preserve my kidney function for the future?

Is there anything I can do to help improve my condition?

Do I feel supported by my healthcare team?

Do I feel supported by my support network?

Am I connected with others with rare kidney diseases or patient support networks?

Who should I contact to receive peer-to peer support?

After your appointment

During or following your appointment, remember to write down:

— Anything that you don't want to forget.

— Set yourself a goal:
Your goal could be related to managing your physical symptoms, emotional symptoms or how you relate to your support network.

Before my next appointment, my goal for managing my condition is:

.....

Follow-up

— If you have additional questions, a member of your healthcare team is often available to assist you afterward.

— Always consult your nephrologist before making any changes to your care plan.

Talk to your healthcare team about any concerns you may have, which could include lifestyle, emotional or other types of issues, such as fertility. This will help them to improve your care. Plus, they may refer you to other healthcare professionals, helpful resources and support groups.

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:

My C3G IC-MPGN
myc3g-icmpgn.com

World Kidney Day
worldkidneyday.org

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.

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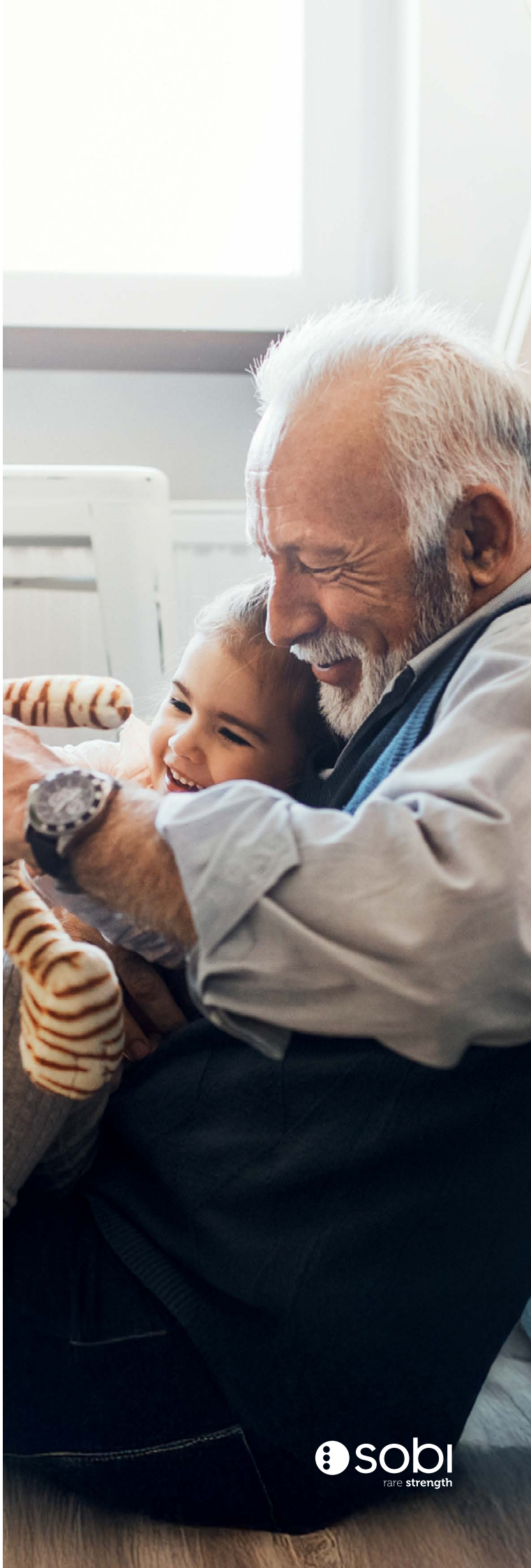
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