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LIVING WITH IgG4-RD

A healthcare appointment guide for patients and providers

Because IgG4-related disease is a relatively recently identified condition, not every physician, nurse, specialist, or emergency care team has heard of it. This guide helps you quickly share what IgG4-RD is, how it has affected your body, and what your healthcare team may need to know to provide effective care. Use it as a starting point for clearer conversations, coordinated care, and shared decision-making.

You may wish to hand the second page to your doctor and keep the first one handy for reference during your visit.

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FOR MY HEALTHCARE TEAM

What caregivers and healthcare providers need to know about IgG4-RD

I have been diagnosed with IgG4-related disease, also called IgG4-RD. IgG4-RD is a systemic immune-mediated disease that can cause inflammation, organ enlargement, and fibrosis in one or more parts of the body. It may present with mass-like lesions that mimic cancer or dysfunction of the affected organs leading to much diagnostic confusion until the full diagnostic picture is reviewed.

IgG4-RD is considered a rare disease and can involve nearly any organ system. Common sites include:

- the **pancreatobiliary system** ("autoimmune pancreatitis" and "IgG4-related sclerosing pancreatitis," which resembles primary sclerosing cholangitis);
- the **major salivary glands** (parotid, submandibular) and the lacrimal glands;
- the **orbits** (extra-ocular muscles);
- the **retroperitoneum** (retroperitoneal fibrosis);
- as well as the **lungs, kidneys, aorta and medium-to-large vessels, lymph nodes**, and other tissues.

Care often involves coordination across rheumatology, gastroenterology, and other specialists, including nephrology and urology, depending on the specific organs involved.

There is no single test that confirms the diagnosis of IgG4-RD in every patient. Diagnosis usually depends on the pattern of symptoms, imaging, blood tests, biopsy findings when available, and careful exclusion of diseases that can mimic it. A normal or mildly elevated serum IgG4 level does not necessarily rule IgG4-RD in or out.

Please consider my known IgG4-RD history when evaluating new symptoms, but also please evaluate urgent symptoms on their own merits. People with IgG4-RD can still have infections, blood clots, heart problems, acute kidney injury, medication side effects, cancer, or other emergencies. They also often have glucose intolerance because of pancreatic involvement by the disease.

Please also be sure to consult my IgG4-RD treating physician before making changes to my IgG4-RD treatment, initiating corticosteroids for another condition, or attributing symptoms to medication side effects.

Important clinical considerations

Because IgG4-RD is rare and can resemble other conditions, a broad differential is often helpful. Please keep in mind:

- A tumor-like lesion may need evaluation for both IgG4-RD and malignancy.
- New symptoms may be related to IgG4-RD, treatment effects, or could be an unrelated acute condition.
- A serum IgG4 measurement can be useful in context (particularly in comparison to historical values, if available), but a high or low serum IgG4 does not confirm or exclude IgG4-RD on its own.
- Before starting or stopping steroids or immune-directed therapy, it may be helpful to consider infection risk, biopsy timing, and input from my treating specialist when feasible.
- Vaccines may need timing around corticosteroids or immune-directed therapies; coordinate with my treating specialist when feasible.

Helpful questions a doctor might ask an IgG4-RD patient

About my diagnosis & disease pattern

- How was IgG4-RD diagnosed?
- Which organs have been affected?
- Which organs are active now?
- Was there a biopsy? Which organ was biopsied?
- Were IgG4 staining or other special pathology tests done?
- Were cancer, infection, lymphoma, or other mimics ruled out?
- What did your first flare look like?
- How does today compare with your usual IgG4-RD symptoms?

About my treatment history

- Which treatments have you used?
- When was your last infusion or steroid course?
- What helped?
- What caused side effects or complications?

About procedures and complications

- Have you had a biliary stent, ureteral stent, nephrostomy tube, port, biopsy, or surgery?
- Have you had bile duct blockage, kidney blockage, pancreatitis, lung disease, eye/orbital disease, vascular involvement, or fibrosis?
- Do you have standing laboratory orders with your IgG4-RD physician?
- How often do you undergo imaging to monitor disease activity?

About follow-up

- Who coordinates you IgG4-RD care?
- Who should be contacted before changing treatment?

References

1. Stone JH, Zen Y, Deshpande V. IgG4-related disease. *New England Journal of Medicine.* 2012;366:539–551.
2. Peyronel F, Della-Torre E, Maritati F, et al. IgG4-related disease and other fibro-inflammatory conditions. *Nature Reviews Rheumatology.* 2025;21:275–290.
3. Arias-Intriago M, Gomolin T, Jaramillo F, et al. IgG4-related disease: Current and future insights into pathological diagnosis. *International Journal of Molecular Sciences.* 2025;26:5325.

Learn more at: IgG4ward! Foundation

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FOR MY HEALTHCARE APPOINTMENT

What I need you to know today

1. **I am here today because:**
2. **My main symptoms right now are:**
3. **These symptoms are:**
   - ☐ New
   - ☐ Getting worse
   - ☐ Similar to a past IgG4-RD flare
   - ☐ Different from my usual IgG4-RD symptoms
   - ☐ Related to a medicine side effect I am worried about
4. **My biggest concern today is:**
5. **What has helped in the past:**
6. **What has not helped or has caused problems:**

Questions I may want to ask during a visit

About today's problem

- What are the most likely causes of my symptoms today?
- Could this be related to IgG4-RD, a medication side effect, an infection, or something unrelated?
- Are there urgent problems we need to rule out today?

About testing

- Do we need blood tests such as CBC, CMP, liver tests, kidney function, inflammatory markers, or IgG subclasses?
- Do we need imaging such as ultrasound, CT, or MRI?
- If a biopsy is being considered, can the pathologist perform stains designed to diagnose IgG4-RD?

About care coordination

- Do you have other patients diagnosed with IgG4-RD?
- Are there other physicians within your practice who are familiar with IgG4-RD?Should my rheumatologist or IgG4-RD specialist be contacted?
- Should any of my current medicines be held, changed, or continued?
- What symptoms should make me seek emergency care after I leave?

About next steps

- What is the plan for follow-up?
- Who will review my test results with me?
- When should I expect results, and whom should I call if I do not hear back?

Red flags that need urgent attention

Please take these symptoms seriously, especially if taking prednisone, rituximab, inebilizumab, or another immune-suppressing medicine.

Seek urgent or emergency care for:

- ☐ Yellow eyes or skin, especially with fever, belly pain, dark urine, or pale stool
- ☐ Fever, chills, confusion, or feeling very ill while immune-suppressed
- ☐ Severe abdominal, back, flank, chest, or head pain
- ☐ Trouble breathing or new chest pain
- ☐ New weakness, numbness, severe headache, vision loss, or eye pain
- ☐ Decreased urination or known kidney/ureter blockage
- ☐ Rapidly growing swelling, lump, or painful gland enlargement
- ☐ Signs of an allergic reaction or serious medication reaction

My IgG4-RD snapshot

My Name:

Date prepared/updated:

My IgG4-RD diagnosis was made in (year):

My affected organs are:

- ☐ Pancreas / bile ducts
- ☐ Salivary glands
- ☐ Tear glands / eyes / orbits
- ☐ Lungs
- ☐ Kidneys
- ☐ Retroperitoneum / Ureters
- ☐ Aorta or blood vessels
- ☐ Lymph nodes
- ☐ Pachymeninges
- ☐ Thyroid gland
- ☐ Other:

My diagnosis was based on:

- ☐ Biopsy
- ☐ Imaging
- ☐ Blood tests
- ☐ Specialist evaluation
- ☐ Other:

My IgG4-RD specialist or main doctor:

Name:

Phone:

Other key Specialist:

Key records my healthcare team may need

Most important records to review or request:

Diagnosis note from my IgG4-RD specialist:

Biopsy/pathology report location:

Imaging reports: CT / MRI / PET-CT / ultrasound:

Most recent blood tests:

Most recent treatment dates:

Implants, stents, ports, drains, or procedures:

Please note: IgG4-RD is usually diagnosed by putting several clues together. **An elevated IgG4 level (via blood test) can be a good indicator, but alone may not prove or rule out the disease.** If there is a new mass, blockage, or organ problem, please evaluate for IgG4-RD and for other urgent causes, including cancer, infection, or obstruction.

My medicines and immune status

Some IgG4-RD therapies modify immune function, which may affect infection risk, vaccine response, wound healing, and medication decisions. Please consider my current and prior treatments when evaluating fever, prescribing steroids or antibiotics, planning procedures, or deciding whether to hold or continue immune-directed therapy.

My current IgG4-RD medicines (dosage/frequency):

Medicines I have taken before for IgG4-RD:

- ☐ Prednisone or other corticosteroids
- ☐ Rituximab / Rituxan
- ☐ Inebilizumab / Uplizna
- ☐ Obexelimab
- ☐ Methotrexate
- ☐ Azathioprine
- ☐ Mycophenolate
- ☐ Other:

Date and type of my last infusion or immune-suppressing treatment:

Important medication issues, side effects, or allergies:

Past treatment complications:

- ☐ Serious infection
- ☐ Low immunoglobulins
- ☐ Infusion reaction / Anaphylaxis
- ☐ Steroid-related diabetes or high blood sugar
- ☐ Bone loss / osteoporosis
- ☐ High blood pressure
- ☐ Mood or sleep problems on steroids
- ☐ Other:

Symptoms that may matter in IgG4-RD

IgG4-RD symptoms depend on which organs are involved. Please tell your healthcare team about symptoms even if they seem unrelated. You may also want to mention symptoms such as fatigue, brain fog, pain, or discomfort that may be part of your day-to-day experience or may worsen during flares.

Swelling or lumps

- ☐ Swelling around the eyes
- ☐ Swelling of salivary glands, jaw, or neck
- ☐ New lump, mass, or enlarged lymph nodes
- ☐ Leg swelling

Breathing or chest symptoms

- ☐ Shortness of breath
- ☐ Cough
- ☐ Chest pain
- ☐ Wheezing

Digestive, pancreas, or bile duct symptoms

- ☐ Yellow eyes or skin
- ☐ Dark urine
- ☐ Pale or clay-colored stool
- ☐ Belly pain
- ☐ Nausea or vomiting
- ☐ Unexplained weight loss
- ☐ New diabetes or blood sugar changes

General symptoms

- ☐ Fever
- ☐ Severe fatigue
- ☐ Night sweats
- ☐ Joint or muscle pain
- ☐ New weakness, numbness, headache, or vision changes

Kidney, ureter, or back symptoms

- ☐ Flank or back pain
- ☐ Trouble urinating
- ☐ Blood in urine
- ☐ Known hydronephrosis or ureter blockage
- ☐ History of ureteral stent
- ☐ Lower extremity swelling (legs and feet)
- ☐ Foamy urine

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