



IgG4ward!

PAVING A PATH 4WARD

Understanding IgG4-related disease (IgG4-RD)

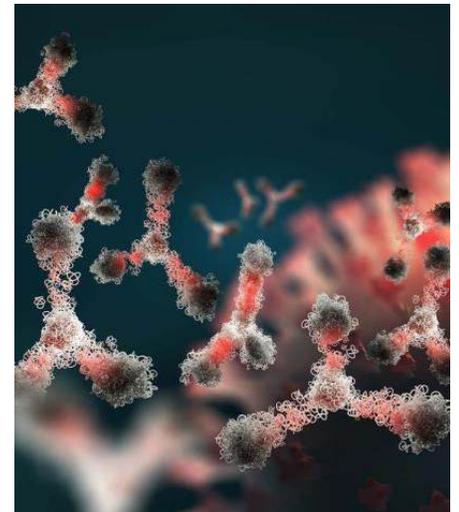
If you or someone you love has been diagnosed with IgG4-related disease (IgG4-RD), a rare and complex condition, you likely have many questions about what this means for your health and future.

IgG4-RD affects each person differently.

For some, symptoms may be mild and manageable, while for others, IgG4-RD can significantly impact daily life and require ongoing medical care.

What is IgG4-related disease?

- **IgG4-Related Disease (IgG4-RD)** is a rare **immune-mediated disorder** – possibly an “autoimmune” disease – but certainly one associated with dysfunction of the immune system. The immune system is the body’s defense system that normally works to prevent infections and fight off infections when they do occur.
- People with IgG4-RD have immune systems that are overly active, leading to inflammation within tissues and scarring that can occur within the organs affected. Many patients, but not all, also have elevated levels of an antibody called **immunoglobulin G4 (IgG4)**, as well. IgG4-RD can impact multiple organ systems, making early diagnosis essential for effective treatment.



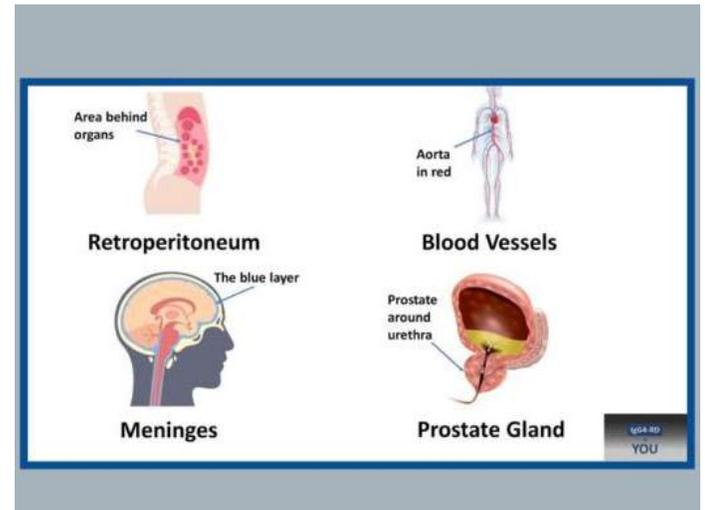
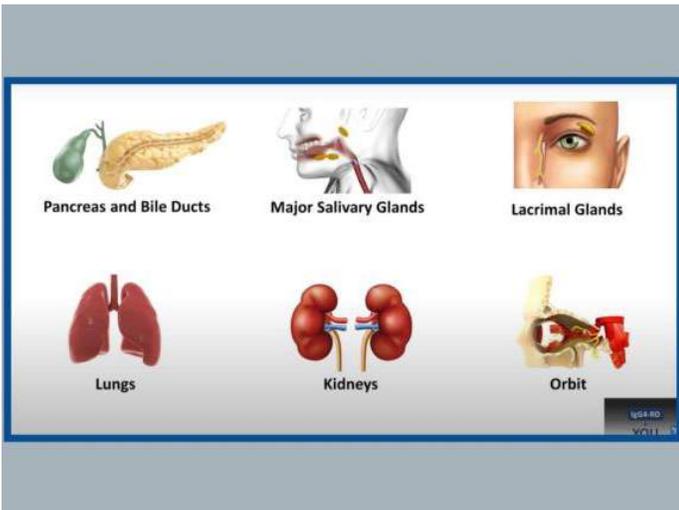
In most IgG4 antibodies, the two halves of the antibody are not identical. This is because while circulating in the blood, the IgG4 antibody literally becomes “unhinged,” that is, the two identical halves disconnect from each other and recombine with another half antibody that is not identical.



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Organ(s) Involved in IgG4-RD:



- Pancreas and Bile Ducts
- Submandibular and Parotid Glands
- Kidneys
- Retroperitoneum
- Meninges
- Lacrimal Glands
- Lungs
- Orbits
- Blood Vessels
- Prostate Gland



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Who is at Risk for Developing IgG4-RD?



IgG4-RD does not discriminate.

The disease can affect people of all ages, sexes and backgrounds. However, it most commonly affects middle- aged to older adults, with a higher prevalence in men. Genetic, environmental and immune system factors may contribute to the development of the disease.

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What are the symptoms of IgG4-related disease?

- Swelling or lumps in affected areas (e.g., neck, face, or abdomen)
- Fatigue and general discomfort
- Pain or discomfort in affected organs
- Dysfunction of involved organs depending on the area



How is IgG4-RD Diagnosed?

Diagnosing IgG4-related disease can be challenging due to its wide range of symptoms and its ability to mimic other conditions.

Physicians rely on a combination of clinical evaluation, imaging and laboratory tests and sometimes pathology studies (biopsies) to confirm an IgG4-related disease diagnosis.

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How is IgG4-RD Diagnosed?

- **Medical History and Physical Examination:**
 - A detailed discussion of symptoms and a physical exam to assess any swelling or organ involvement.
- **Blood Tests:**
 - IgG4, IgG1, IgE, C3, C4
- **Imaging Studies that may include:**
 - A computed tomography (CT) scan, commonly referred to as a "CAT scan,"
 - Magnetic Resonance Imaging (MRI)
 - Ultrasound
 - Positron Emission Tomography (PET) Scan
- **Biopsy of Affected Tissue:**
 - A biopsy is often necessary to confirm the diagnosis, revealing inflammation, fibrosis, and IgG4-positive plasma cells in the tissue.
- **Additional Laboratory Tests:**
 - Lipase, creatinine, hemoglobin A1c, elastase measurement (in stool)
- **Organ function tests** may be conducted based on which organs are affected. For example, in patients with active pancreatic inflammation, the blood lipase level is often high. In a person with a severely damaged pancreas, the lipase level is often very low.

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Do Doctors use guidelines to determine a diagnosis?

Doctors use specific guidelines to classify a patient as having IgG4-RD.

The *American College of Rheumatology (ACR)* and the *European League Against Rheumatism (EULAR)* created these guidelines in 2019 to help doctors worldwide.

These criteria were intended primarily for research studies but can also provide reassurance in clinical situations that the diagnosis is correct.

Some patients have a correct diagnosis of IgG4-RD but might not fulfill the ACR/EULAR Criteria. (This is often because the patient has not had a biopsy).

Biopsies are not always required to be confident of an IgG4-RD diagnosis, but sometimes they are essential confirming the clinical suspicion or eliminating mimickers of IgG4-RD.

For more details on the ACR/EULAR guidelines:
Visit the [official ACR website](#) or the [EULAR website](#).

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What Kind of Doctors Generally Treat IgG4-RD?

IgG4-RD often requires a multidisciplinary approach. Specialists who commonly manage this disease include:



- Rheumatologists (immune system disorders)
- Gastroenterologists (digestive system involvement)
- Endocrinologists (diabetes resulting from pancreatic damage)
- Allergists (symptoms of asthma or allergy)
- Pulmonologists (lung involvement)
- Nephrologists (kidney involvement)
- Hematologists (high levels of immunoglobulins - antibodies - in the blood)

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How is IgG4-RD Treated?

While there is no cure for IgG4-related disease, treatment focuses on managing symptoms, reducing inflammation and preventing organ damage.

With the right treatment plan, individuals with IgG4-related disease can maintain a good quality of life. Treating IgG4-RD is a team effort – so you and your doctors will work together to find the treatment plan that's right for you.



What are the common treatments for IgG4-RD?

The treatment options available for IgG4-RD, including:

- **Glucocorticoids:** Often the first line of treatment to quickly reduce inflammation. Prednisone is commonly prescribed.
- **Immunosuppressive Medications:** Drugs such as azathioprine or mycophenolate mofetil may be used to suppress the immune system and prevent disease progression.
- **Biologic Therapies:** a medication that targets specific immune cells.

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Why is regular monitoring important for IgG4-RD patients?

- Regular monitoring through blood tests and imaging studies is essential to assess disease activity and adjust treatment as needed. Monitoring helps to ensure the effectiveness of the treatment plan and to detect any potential complications early.

What are the potential side effects of IgG4-RD treatments?

- All medications used to treat IgG4-RD may have side effects, which may include:
 - lowered immunity, increased risk of infections, fatigue, weight gain, high blood pressure, mood changes and gastrointestinal discomfort.
- The extent and severity of side effects vary from person to person and depend on factors such as dosage, duration of treatment, individual health status and the specific medication being used.
 - For example, B-cell depletion therapies may increase susceptibility to infections and cause infusion reactions, while glucocorticoids may lead to osteoporosis, muscle weakness and blood sugar imbalances.
- Patients should work closely with their healthcare providers to manage potential side effects, adjust treatment plans as needed and take preventive measures such as vaccinations, bone health support and lifestyle modifications.
- Open communication about any new or worsening symptoms can help optimize care and improve overall well-being.



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Is surgery ever required for IgG4-RD?

- Surgical intervention may be necessary to address complications such as organ damage or obstruction caused by the disease.

How can I reduce my risk of infections while on treatment?

- Infection prevention is critical when undergoing treatment for IgG4-RD. It is important to stay up to date with vaccinations, such as:
 - Influenza vaccine
 - Pneumonia vaccine
 - COVID-19 vaccine
 - Shingles vaccine
 - RSV vaccine
- Discuss with your healthcare provider to ensure you receive the appropriate vaccinations based on your treatment plan.

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Medical Follow-up and Flares:



Even with effective treatment, flares of IgG4-RD are possible. Regular doctor visits and ongoing monitoring with blood tests and imaging studies are essential to detect and address flares early. Open communication with your healthcare team can help you manage any returning symptoms or changes in your condition.

If you need help finding a physician in your area that treats IgG4-RD - please visit the [IgG4ward! Physician's Network](#)

Thriving with IgG4-RD

Managing IgG4-related disease is about more than just medical treatment.

It's important to:

- **Stay informed:** Learn about your condition and treatment options
- **Work closely with your healthcare team:** Regular check-ins and open communication are vital
- **Prioritize self-care:** Balanced nutrition, physical activity, and stress management can support overall well-being
- **Seek support:** Connecting with others through support groups or the IgG4ward! Foundation can help you feel less alone

