

UNDERSTANDING IgG4-RD

From difficult symptoms to an accurate diagnosis to effective treatment, the rare disease journey is a long one. But despite the challenges patients face, innovation and education are regularly improving lives.

For a rare condition known as IgG4-RD, increased education is needed to help patients better understand their disease. Updated resources can also inform health care providers about new treatments and best practices for managing the disease.



Q: What is IgG4-RD?

IgG4-related disease, frequently called IgG4-RD, is a rare and chronic autoimmune disease.

This rare condition often manifests as tumors in affected organs and can impact multiple systems in the body. Individual patients may experience the disease in vastly different ways.

This rare condition is a fairly recent discovery, often associated with autoimmune pancreatitis. As a result, few providers have a firm grasp on the condition, and patients may experience misdiagnosis or a lack of diagnosis. When patients do receive a proper diagnosis, however, they can work with their providers to develop treatment plans to best manage their condition.

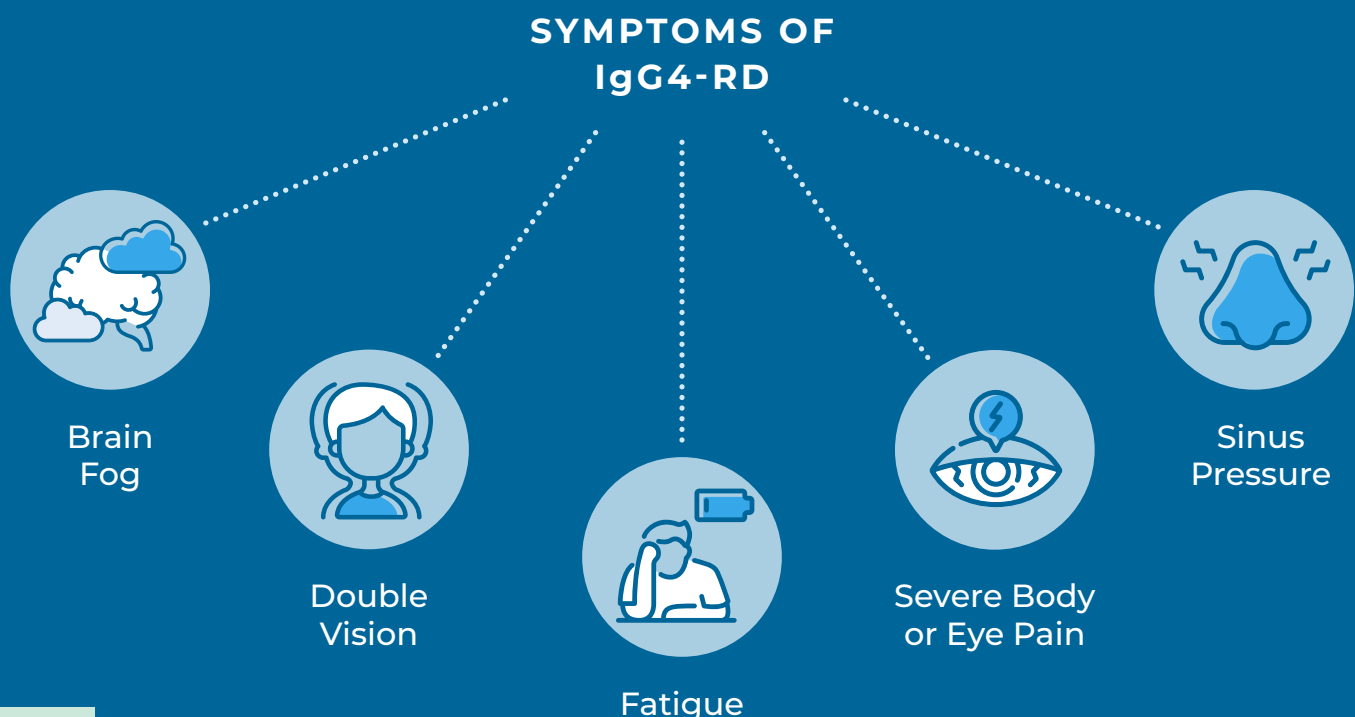
Q: What are the symptoms?

Symptoms may vary from patient to patient. They may include, but are not limited to, any combination of:

- **Brain fog**
- **Double vision**
- **Fatigue**
- **Severe body or eye pain**
- **Sinus pressure**

Symptoms vary based on the organs affected by IgG4-RD.

Some patients may not experience any symptoms for months or years before they receive a diagnosis, resulting in serious organ damage.



Catching IgG4-RD early can improve the effectiveness of treatment.



Q: How do patients get a diagnosis?

IgG4-RD can prove challenging to diagnose because symptoms resemble those of other conditions, such as cancer. As patients search for a proper diagnosis, they may spend time and money seeking out specialist after specialist. Meanwhile, they continue to suffer from painful and debilitating symptoms.

Comorbidities can also complicate the diagnosis process, as determining which symptoms correspond to which disease may lead providers to an incorrect conclusion. Some patients may even receive an incorrect diagnosis, resulting in treatment that might worsen their condition.

Biopsies can help providers better identify what is occurring in a patient's body. Although some patients may have elevated levels of IgG4, this may not be the case for all.

Q: How is IgG4-RD treated?

While no FDA-approved treatments specifically for IgG4-RD patients are available yet, health care providers can develop personalized treatment regimens for patients. These approaches often consist of medications approved for other diseases and used off label for IgG4-RD.

Treatments may vary based on the impacted organs, a patient's individual symptoms and IgG4-RD's progression in the body. Patients and providers work not only to address the condition but also to avoid relapse once a patient is in remission.

Catching IgG4-RD early can improve the effectiveness of treatment, as IgG4-RD responds better to early treatment.

Q: What types of providers treat IgG4-RD?

The location of IgG4-RD, as well as which organs and systems are impacted, can influence what type of provider a patient sees for treatment initially. Most patients end up seeing rheumatologists, who generally have the most expertise in this area. Patients may, however, also be seen by different types of health care professionals, such as:

- **Emergency medical specialists**
- **Gastroenterologists**
- **Nephrologists**
- **Neurologists**
- **Ophthalmologists**
- **Pathologists**

There are few IgG4-RD specialists, making providers for this patient community a large unmet need.

Still, many IgG4-RD patients turn to the specialists who are familiar with the disease to determine diagnosis and treatment. These providers understand the multisystemic nature of the disease and can best address it. Having a patient-provider relationship built on trust also plays an important role, as symptoms can vastly differ and be severe.

Q: Are more treatments in development?

Several clinical trials are currently underway and searching for participants. As more clinical research on IgG4-RD is completed, researchers will gain a better understanding of the causes and impacts of this rare condition. As a result, innovative treatments will be able to help patients treat and manage their condition.

However, patients may experience barriers when trying to participate in clinical trials, such as:

- **Knowledge of trials and eligibility**
- **Geographical distance from the trial site**
- **Travel-associated expenses**

Addressing these barriers is critical to ensuring new treatments may become available.

IgG4-RD patients can work with their health care provider to identify the right trial to participate in.



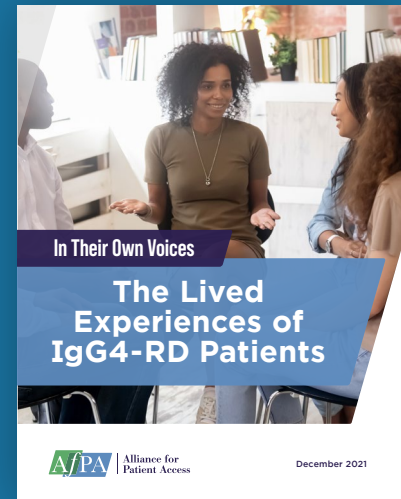


LEARN MORE

Learn more about potential clinical trials opportunities for people living with IgG4-RD at ClinicalTrials.gov.

Read more about IgG4-RD from patients in [The Lived Experiences of IgG4-RD Patients](#) and the [report update](#).

The Alliance for Patient Access' Rare Diseases Working Group has launched a clinician advocacy initiative on IgG4-RD. To get involved, contact Elizabeth Simpson at esimpson@allianceforpatientaccess.org.



RESOURCES

1. IgG4-Related Disease. American College of Rheumatology. <https://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/IgG4-Related-Disease-IgG4-RD>.
2. Legatowicz-Koprowska M. IgG4-related disease: why is it so important?. *Cent Eur J Immunol*. 2018;43(2):204-208. doi:10.5114/ceji.2018.77391
3. Nirula A, Glaser SM, Kalled SL, Taylor FR. What is IgG4? A review of the biology of a unique immunoglobulin subtype [published correction appears in *Curr Opin Rheumatol*. 2011 Mar;23(2):227. Taylor, Frederick R [corrected to Taylor, Frederick R]]. *Curr Opin Rheumatol*. 2011;23(1):119-124. doi:10.1097/BOR.0b013e3283412fd4



The Alliance for Patient Access is a national network of policy-minded health care providers advocating for patient-centered care.

To learn more visit AllianceforPatientAccess.org