



PATIENT FACT SHEET

IgG4-Related Disease (IgG4-RD)



CONDITION DESCRIPTION

IgG4-related disease (IgG4-RD) is an immune-mediated condition, meaning that it involves the occurrence of disease in organs as the result of an abnormally regulated immune system. IgG4-RD is an autoimmune condition much like rheumatoid arthritis and systemic lupus erythematosus. The classic IgG4-RD patient is a middle-aged to elderly individual – more likely to be male than female. In rare cases, the disease also affects children.

Some patients with IgG4-RD have disease in only one organ. Others, however, have disease that affects multiple organs at the same time. It is not unusual for disease to affect anywhere between two to six organs, and sometimes more, in a single patient.

The following organs are often involved in IgG4-RD: (1) **meninges** (layer of tissues surrounding the brain and spinal cord), (2) **orbits** (eye sockets), (3) **lacrimal glands** (upper outer portion of the eyes that produce tears), (4) **major salivary glands** (glands that produce saliva to aid in the digestion of food), (5) **thyroid gland** (gland sits low on the front of the neck and produces hormone that helps the regulation of body metabolism), (6) **lungs**, (7) **aorta** (the body's largest blood vessel that supplies oxygenated blood to the circulatory system), (9) **kidneys** (10) **pancreas** (organ in abdomen), (11) **bile ducts** (located both within and outside the liver).



SIGNS/ SYMPTOMS

Many patients with IgG4-RD have no signs or symptoms for months or even years before the diagnosis is made, which can cause organ damage even while the patient is feeling well, long before he or she comes to medical attention. Because of the many organs that IgG4-RD can affect, the disease can exist in multiple ways associated with many symptoms. IgG4-RD can often appear as a mass that mimics a cancer. It can also often be associated with allergic symptoms (e.g., asthma, allergic rhinitis, eczema, etc.), which may not be linked to IgG4-RD for years because these symptoms are so common in the general population.

Other common symptoms include fatigue, weight loss, headaches, dysfunction of the cranial nerves (control eye movements, speech, swallowing, hearing, and other critical functions), bulging of one or both eyes, bulges on the sides of the face or below the chin, inflammatory tissue in the thyroid, large-vessel vasculitis (inflammation in the blood vessel wall), shortness of breath, blockage of urine flow from the kidney, enlarged kidneys, abdominal pain and painless jaundice (yellow tint to the skin or eyes).



COMMON TREATMENTS

If diagnosed before serious organ damage has occurred, IgG4-RD typically responds well to treatment, but chronic therapy is necessary. Glucocorticoids are typically viewed as the initial treatment of IgG4-RD. Although nearly all patients with IgG4-RD respond to glucocorticoids, approximately 40% of those fail to

achieve complete remission or relapse within one year. The disease often recurs after prednisone tapering, as well. Rituximab, however, is frequently a good treatment for IgG4-RD and it is not typically associated with many of the adverse effects linked to glucocorticoids.



CARE/ MANAGEMENT TIPS

Patients with IgG4-RD need to have close follow-ups with physicians who are knowledgeable about IgG4-RD. They also need to broaden their own knowledge of this condition and be aware of the symptoms and complications

that can result from disease in the organs. Because of the organ damage IgG4-RD, it is important to seek medical attention for symptoms early and begin treatment.