ANTI-GBM/GOODPASTURE'S SYNDROME

What is anti-GBM/Goodpasture's syndrome?

Anti-GBM/Goodpasture's syndrome is a rare, but serious autoimmune disease that causes inflammation of the kidneys and lungs. In this syndrome, the immune system mistakenly makes "anti-GBM antibodies" which attack the lungs and kidneys, causing bleeding within the organs. The disease can progress rapidly to life-threatening organ failure, so early diagnosis and treatment are vital. Smoking and exposure to certain chemicals may be among the triggers for this disease.

Anti-GBM (anti-glomerular basement membrane disease) and Goodpasture's syndrome are frequently discussed together. Anti-GBM disease may affect only the kidneys, however when it causes both kidney and lung disease, it is called Goodpasture's syndrome. This disease may come on suddenly, with early symptoms of fatigue, nausea, pale skin and shortness of breath. As the disease progresses, patients may cough up blood, or have bloody urine, leg swelling, and chest pain, among other signs and symptoms.

Treatment for anti-GBM/Goodpasture's syndrome typically includes corticosteroids such as prednisone, and medications that suppress the immune system. Most patients also undergo a plasma exchange procedure, which removes the harmful antibodies from the blood. Anti-GBM/Goodpasture's syndrome can last up to two years. During this time, ongoing medical care is essential to monitor disease progression, medication side effects and potential relapses.

Causes

The cause of anti-GBM/Goodpasture's syndrome is not fully understood by researchers. It is thought to be an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissue. Researchers believe a combination of factors may trigger anti-GBM/Goodpasture's syndrome including genetics, respiratory infections, exposure to certain chemicals (hydrocarbon fumes, metallic dust), use of certain drugs such cocaine, and tobacco smoking.

Who gets anti-GBM/Goodpasture's syndrome?

Anti-GBM/Goodpasture's syndrome is rare, with an estimated incidence of about 1 case per million. It typically affects people in two age groups, young people aged 20 to 30, and people 60 and older. It is most common among young Caucasian men, and is rare in children.

Symptoms

Early symptoms include general body aches and pains, fatigue, weakness or lethargy, pale skin, nausea, lack of appetite, and shortness of breath. Anti-GBM/Goodpasture's syndrome can progress rapidly to the lungs and kidneys, with the following symptoms:

- Persistent dry cough
- Coughing up blood (Bleeding in the lungs can cause respiratory failure, so seek immediate medical attention if you are coughing up blood.)
- Bloody and/or foamy urine
- Difficult or painful urination
- Swelling in the legs, hands or feet
- Chest pain

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Complications

Both kidney and lung failure are life-threatening complications of anti-GBM/Goodpasture's syndrome. Kidney failure is the most serious complication, and may require kidney dialysis or even kidney transplant. The disease can also cause life-threatening bleeding in the lungs, however early diagnosis and treatment can help prevent long-term lung damage.

Diagnosis

There is no single test to diagnose anti-GBM/Goodpasture's syndrome. Your doctor will consider a number of factors, including a detailed medical history; physical examination; laboratory tests; and specialized imaging studies. Diagnosis is usually confirmed by the presence of anti-GBM antibodies in a blood test, and through a kidney biopsy—the gold standard for diagnosis of this disease.

Diagnosis involves ruling out diseases that cause similar symptoms, including other forms of vasculitis. Depending on organs affected, the following diagnostic tests may be ordered:

- **Urinalysis:** Excessive protein or presence of red blood cells may indicate kidney inflammation.
- **Blood tests:** Detecting the presence of anti-GBM antibodies in the bloodstream is key to confirming a diagnosis of anti-GBM/Goodpasture's syndrome.
- **Imaging studies:** Chest X-ray or computed tomography (CT) scan may reveal bleeding or lung changes/damage.
- **Tissue biopsy:** This surgical procedure removes a small tissue sample from the kidneys or lungs, which is examined under a microscope for signs of inflammation or tissue damage.

Treatment

Treatment for anti-GBM/Goodpasture's syndrome is aimed at reducing inflammation, halting further antibody production, and removing anti-GBM antibodies from the bloodstream. Medications include corticosteroids such as prednisone, and the immunosuppressant medication cyclophosphamide. Intravenous corticosteroids may be required to control bleeding in the lungs.

Treatment also usually requires plasmapheresis (plasma exchange), in which the liquid part of the blood (plasma) is removed, filtered of the harmful anti-GBM antibodies, and then returned to the body. This procedure is typically done daily for several weeks. If the disease leads to kidney failure, kidney dialysis or transplant will likely be required.

Side effects

The medications used to treat anti-GBM/Goodpasture's syndrome have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. It's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

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Relapse

Relapses are rare with anti-GBM/Goodpasture's syndrome. However, if your symptoms return, or you develop new ones, report them to your doctor as soon as possible.

Your medical team

Effective treatment of anti-GBM/Goodpasture's syndrome may require a team of medical providers and specialists. In addition to your primary care provider, you may need to see the following specialists: a rheumatologist (joints, muscles, immune system); nephrologist (kidneys); pulmonologist (lungs); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track your medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member if necessary to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, be sure to speak with your medical team. It is always your right to seek a second opinion.

Living with anti-GBM/Goodpasture's syndrome

Living with a chronic disease can be challenging at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

Anti-GBM/Goodpasture's syndrome requires early and aggressive treatment to avoid life-threatening lung and kidney failure. The outlook especially depends on the disease's effect on the kidneys. Patients have the best outcome when treatment is started before kidney dialysis is required. Relapse is uncommon with this syndrome and it doesn't typically require long-term treatment. Avoiding cigarette smoke and inhaled toxins may improve a patient's outlook.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

For more information on vasculitis research, visit: www.vasculitisfoundation.org/research

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About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

About the Vasculitis Foundation

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

Connect with the VF on Social Media

- Instagram: vasculitisfoundation
- Twitter: @VasculitisFound
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

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Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

Join the VPPRN!

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

Vasculitis Foundation

PO Box 28660
Kansas City, Missouri, 64188-8660
USA
Phone: 816.436.8211
Toll-free: 800.277.9474
Fax: 816.656.3838
www.VasculitisFoundation.org
Email: vf@vasculitisfoundation.org

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