

What is anti-GBM/Goodpasture's syndrome?

Anti-GBM/Goodpasture's syndrome (syndrome means a group of symptoms) is a rare condition that causes inflammation of the kidneys and lungs.

Anti-glomerular basement membrane (anti-GBM) disease may affect only the kidneys; however, when it causes both kidney and lung disease, it is called Goodpasture's syndrome.

In this syndrome, the immune system mistakenly makes "anti-GBM antibodies," which attack the lungs and kidneys. This causes bleeding in these organs. The condition can cause the organ to die. Early diagnosis and treatment are important to prevent this.

Causes

The causes of anti-GBM/Goodpasture's syndrome are not fully understood. It is thought to be an autoimmune disorder, which is a condition that occurs when the body's natural defense system mistakenly attacks healthy tissue.

It is believed that a combination of factors may trigger anti-GBM/Goodpasture's syndrome:

- Genetics
- Respiratory infections
- Exposure to chemicals, such as:
 - hydrocarbon fumes, metallic dust
 - use of certain drugs such cocaine, and tobacco smoking

Symptoms

Early symptoms include:

- General body aches and pains
- Fatigue
- Weakness or lethargy
- Pale skin
- Nausea
- Lack of appetite
- 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; seek immediate medical attention if you are coughing up blood)
- Chest pain
- Bloody and/or foamy urine
- Difficult or painful urination
- Swelling in the legs, hands and/or feet

Complications

Both kidney and lung failure are life-threatening complications of anti-GBM/Goodpasture's syndrome. Kidney failure may require kidney dialysis or even a 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

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:** These are used to detect the presence of anti-GBM antibodies, check blood cell counts and kidney function.
- **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 or stopping further antibody production
 - Medications include corticosteroids such as prednisone and the immunosuppressant medication cyclophosphamide. Intravenous corticosteroids may be required to control bleeding in the lungs.
- Removing anti-GBM antibodies from the bloodstream

- Plasmapheresis (plasma exchange), in which the liquid part of the blood (plasma) is removed, the harmful anti-GBM antibodies are removed and then plasma is returned to the body. This procedure is typically done for one to two weeks.

If the disease leads to kidney failure, kidney dialysis or transplant will likely be required.

Side Effects of Medications

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
- 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.

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.

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.

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:

- Rheumatologist (joints, muscles, immune system)
- Nephrologist (kidneys)
- Pulmonologist (lungs)
- Others as needed

The best way to manage your disease is to be an active partner with your health care providers and to 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 severity and how soon treatment is started.

Relapse is uncommon with this syndrome and it doesn't typically require long-term treatment.

Avoiding cigarette smoke and inhaled toxins may improve your outlook.

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