

Goodpasture Syndrome

What is Goodpasture syndrome?

Goodpasture syndrome is a pulmonary-renal syndrome, which is a group of acute illnesses involving the kidneys and lungs. Goodpasture syndrome includes all of the following conditions:

- glomerulonephritis—inflammation of the glomeruli, which are tiny clusters of looping blood vessels in the kidneys that help filter wastes and extra water from the blood
- the presence of anti-glomerular basement membrane (GBM) antibodies; the GBM is part of the glomeruli and is composed of collagen and other proteins
- bleeding in the lungs

In Goodpasture syndrome, immune cells produce antibodies against a specific region of collagen. The antibodies attack the collagen in the lungs and kidneys.

Ernest Goodpasture first described the syndrome during the influenza pandemic of 1919 when he reported on a patient who died from bleeding in the lungs and kidney failure. Diagnostic tools to confirm Goodpasture syndrome were not available at that time, so it is not known whether the patient had true Goodpasture syndrome or vasculitis. Vasculitis is an autoimmune condition—a disorder in which the body's immune system attacks the body's own cells and organs—that involves inflammation in the blood vessels and can cause similar lung and kidney problems.

Goodpasture syndrome is sometimes called anti-GBM disease. However, anti-GBM disease is only one cause of pulmonary-renal syndromes, including Goodpasture syndrome.

Goodpasture syndrome is fatal unless quickly diagnosed and treated.

What causes Goodpasture syndrome?

The causes of Goodpasture syndrome are not fully understood. People who smoke or use hair dyes appear to be at increased risk for this condition. Exposure to hydrocarbon fumes, metallic dust, and certain drugs, such as cocaine, may also raise a person's risk. Genetics may also play a part, as a small number of cases have been reported in more than one family member.

What are the symptoms of Goodpasture syndrome?

The symptoms of Goodpasture syndrome may initially include fatigue, nausea, vomiting, and weakness. The lungs are usually affected before or at the same time as the kidneys, and symptoms can include shortness of breath and coughing, sometimes with blood. The progression from initial symptoms to the lungs being affected may be very rapid. Symptoms that occur when the kidneys are affected include blood in the urine or foamy urine, swelling in the legs, and high blood pressure.

How is Goodpasture syndrome diagnosed?

A health care provider may order the following tests to diagnose Goodpasture syndrome:

- **Urinalysis.** Urinalysis is testing of a urine sample. The urine sample is collected in a special container in a health care provider's office or commercial facility and can be tested in the same location or sent to a lab for analysis. For the test, a nurse or technician places a strip

of chemically treated paper, called a dipstick, into the urine. Patches on the dipstick change color when protein or blood are present in urine. A high number of red blood cells and high levels of protein in the urine indicate kidney damage.

- **Blood test.** A blood test involves drawing blood at a health care provider's office or commercial facility and sending the sample to a lab for analysis. The blood test can show the presence of anti-GBM antibodies.
- **Chest x ray.** An x ray of the chest is performed in a health care provider's office, outpatient center, or hospital by an x-ray technician, and the images are interpreted by a radiologist—a doctor who specializes in medical imaging. Abnormalities in the lungs, if present, can be seen on the x ray.
- **Biopsy.** A biopsy is a procedure that involves taking a piece of kidney tissue for examination with a microscope. The biopsy is performed by a health care provider in a hospital with light sedation and local anesthetic. The health care provider uses imaging techniques such as ultrasound or a computerized tomography scan to guide the biopsy needle into the kidney. The tissue is examined in a lab by a pathologist—a doctor who specializes in diagnosing diseases. The test can show crescent-shaped changes in the glomeruli and lines of antibodies attached to the GBM.

How is Goodpasture syndrome treated?

Goodpasture syndrome is usually treated with

- **immunosuppressive medications**, such as cyclophosphamide, to keep the immune system from making antibodies
- **corticosteroid medications** to suppress the body's autoimmune response
- **plasmapheresis**—a procedure that uses a machine to remove blood from the body, separate certain cells from the plasma, and return just the cells to the person's body; the anti-GBM antibodies remain in the plasma and are not returned to the person's body

Plasmapheresis is usually continued for several weeks, and immunosuppressive medications may be given for 6 to 12 months, depending on the response to therapy. In most cases, bleeding in the lungs stops and no permanent lung damage occurs. Damage to the kidneys, however, may be long lasting. If the kidneys fail, blood-filtering treatments called dialysis or kidney transplantation may become necessary.

Eating, Diet, and Nutrition

Eating, diet, and nutrition have not been shown to play a role in causing or preventing Goodpasture syndrome.

Points to Remember

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 - bleeding in the lungs

- Goodpasture syndrome is fatal unless quickly diagnosed and treated.
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- A urinalysis, blood test, chest x ray, and kidney biopsy are used to diagnose Goodpasture syndrome.
- Goodpasture syndrome is usually treated with immunosuppressive medications, corticosteroid medications, and plasmapheresis.

Reference:

<http://kidney.niddk.nih.gov>

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