Cogan's Syndrome

What is Cogan's Syndrome?

Cogan's syndrome is a rare autoimmune disease that primarily affects the eyes and inner ears, but can also cause inflammation of the blood vessels (vasculitis). Cogan's syndrome is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Cogan's syndrome primarily affects the large vessels, especially the aorta, which carries oxygenated blood from the heart to the rest of the body.

The most common symptoms of Cogan's syndrome include eye pain and redness, excess tear production, vision impairment, loss of hearing, and dizziness. Associated vasculitis symptoms may include joint and muscle pain, along with heart problems such as congestive heart failure.

Cogan's syndrome is treated with medications that suppress the immune system including corticosteroids to control inflammation. In more advanced cases, surgical procedures on the ears, eyes, or even the heart, may be necessary. Early diagnosis and treatment are important to lower the risks of permanent hearing or vision loss. Cogan's syndrome is a chronic disease with periods of relapse and remission, so ongoing medical care is necessary.

Causes

The exact cause of Cogan's syndrome is not fully understood. It is thought to be an autoimmune disorder—a disease that occurs when the body's natural defense system mistakenly attacks healthy tissue. For some people, Cogan's syndrome develops after an infection.

Who Gets Cogan's Syndrome?

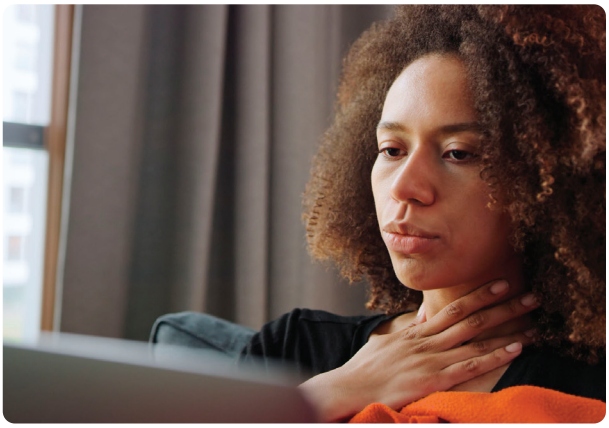
Cogan's syndrome can occur in people of any age, but most frequently affects young adults in their 20s and 30s. It is very rare. The disease can also occur in people of any race, but is more common in Caucasians.

Symptoms

Cogan's syndrome typically starts with either eye inflammation or inner ear inflammation, but often progresses to include both. Vasculitis symptoms can be present at the onset of Cogan's syndrome, or may develop later in the course of the disease. In some cases, symptoms come and go.

The most common symptoms of Cogan's syndrome include:

- Eye redness and pain, decreased or blurred vision, sensitivity to light, and excess tear production
- Hearing loss, in some cases permanent, accompanied by a sensation of pressure in the ear and/or ringing in the ears (tinnitus)
- Vertigo (a sense of the room spinning) and general dizziness
- Poor balance
- Vasculitis symptoms include muscle pain and cramping, joint pain, headache, fever, and weight loss. Heart murmurs or other heart problems may develop.



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Complications

Permanent hearing loss is common in patients with Cogan's syndrome. Vertigo is more severe with initial episodes, tending to improve with time, although poor balance can be persistent. Permanent eye damage and vision loss are uncommon. Vasculitis that affects the aorta can lead to congestive heart failure. Life-threatening complications of Cogan's syndrome are rare, but may include an aortic aneurysm— an abnormal bulge in a weakened artery wall that can rupture.

Diagnosis

There is no single test for diagnosing Cogan's syndrome, so your doctor will consider several factors including a detailed medical history, physical examination, laboratory tests, specialized imaging studies, and biopsy, when indicated.

Your doctor will attempt to rule out diseases with similar symptoms. These include infections, inflammatory disorders such Crohn's disease, other vasculitic syndromes, rheumatoid arthritis, and systemic lupus, certain cancers, and multiple sclerosis, among others. In addition, blood tests and urinalyses will likely be ordered.

The diagnosis of vasculitis is usually confirmed in patients with Cogan's syndrome via:

- Detailed exam by an eye doctor (ophthalmologist) and an ear, nose, and throat specialist (ENT)
- Echocardiography—a cardiac ultrasound
- Magnetic resonance angiography (MRA) for evaluation of blood vessels
- A biopsy of affected tissue—surgical removal and analysis of a small tissue sample from a blood vessel or affected organ

Treatment

Treatment of Cogan's syndrome depends on symptoms, severity of disease, and whether vasculitis is present. Corticosteroid medications are typically started early in the course of Cogan's syndrome for eye inflammation and decreased hearing.

Mild eye disease may initially be treated with topical steroids and nonsteroidal anti-inflammatory drugs (NSAIDs). For more severe disease, oral corticosteroids such as prednisone, and other immunosuppressive medications may be prescribed including methotrexate, cyclophosphamide, cyclosporine, mycophenolate mofetil or azathioprine.

If hearing is impaired and does not respond to medications, cochlear implants may help improve hearing. Cochlear implants are surgically implanted electronic devices that provide a sense of sound to those who are deaf or hard of hearing. Medications may also be prescribed to treat balance problems.

When the cornea (the transparent layer forming the front of the eye) has been severely damaged by inflammation, corneal transplants may be an option. A corneal transplant is a surgical procedure that replaces the scarred cornea with another from an organ donor.

If inflammation of the aorta and/or other vessels are proven to be present, treatments include corticosteroids with other immunosuppressive therapy. Heart problems may require surgical procedures such as aortic valve replacement.

Side Effects of Treatment

The medications used to treat Cogan's syndrome have potentially serious side effects such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, 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 vaccines (*e.g.,* flu shot, pneumonia and/or shingles vaccination), which can reduce your risk of infection.

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Medical Follow-up/Relapse

Cogan's syndrome is a chronic disease with periods of relapse and remission. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular checkups and ongoing monitoring of laboratory and imaging tests are important in detecting relapses early.

Your Medical Team

Effective treatment of Cogan's syndrome may require a team of medical providers. In addition to a primary care doctor, Cogan's syndrome patients will likely need to see the following specialists:

- Ophthalmologist (eyes)
- Otolaryngologist (ear, nose and throat)
- Rheumatologist (joints, muscles, and immune system)
- Cardiologist (heart) 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 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 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 about your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with Cogan's Syndrome

Living with a chronic condition such as Cogan's syndrome can be overwhelming 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

Most patients with Cogan's syndrome respond well to treatment; however, in some cases, the disorder can lead to permanent hearing or vision loss. Early diagnosis and treatment can help minimize these risks. The long-term outlook also depends on whether vasculitis is present and its severity. Cogan's syndrome is a chronic disease, so ongoing medical care is important.

In 2021 the American College of Rheumatology (ACR) published guidelines for the management of certain vasculitides, which were also endorsed by the Vasculitis Foundation (VF). Clinical practice guidelines are developed to reduce inappropriate care, minimize geographic variations in practice patterns, and enable effective use of health care resources. Guidelines and recommendations developed and/or endorsed by the ACR are intended to provide guidance for particular patterns of practice and not to dictate the care of a particular patient. The application of these guidelines should be made by the physician in light of each patient's individual circumstances. Guidelines and recommendations are subject to periodic revision as warranted by the evolution of medical knowledge, technology, and practice.

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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 disease (formerly Goodpasture's syndrome)
- Aortitis
- Behçet's syndrome
- Central nervous system vasculitis (CNSV)
- Cogan's syndrome
- Cryoglobulinemic vasculitis
- Cutaneous small-vessel vasculitis (CSVV) (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis (GCA)
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (formerly Henoch-Schönlein purpura)
- Kawasaki disease
- Microscopic polyangiitis (MPA)
- Polyarteritis nodosa (PAN)
- Polymyalgia rheumatica (PMR)
- Rheumatoid vasculitis
- Takayasu arteritis (TAK)
- Urticarial vasculitis (normocomplementemic or hypocomplementemic)

About the VF

The VF is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF is a 501(c)(3) nonprofit organization governed by a Board of Directors and advised on medical issues by a Medical and Scientific Advisory Board. VF's educational materials are not intended to replace the counsel of a physician. The VF does not endorse any medications, products, or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

The VF gratefully acknowledges Alexandra Villa-Forte, MD, Cleveland Clinic Center for Vasculitis Care and Research, for her expertise and contribution to this brochure.

To access additional VF support and educational resources, please scan the QR code below.

VF 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.

VASCULITIS FOUNDATION®

Early Diagnosis · Better Treatment · Improving Lives

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