

## 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—a condition known as vasculitis. Vasculitis is a family of rare disorders characterized by narrowing of the vessels, which restricts blood flow and damages 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 initially treated with corticosteroids such as prednisone, along with medications that suppress the immune system to control inflammation. In more advanced cases, surgical procedures on the eyes, ears, 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 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. For some people, Cogan's syndrome develops after an infection. It is not known to run in families.

### 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. 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 syndrome. 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.

## COGAN'S SYNDROME

### 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 visual 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 a number of factors, including a detailed medical history; physical examination; laboratory tests; specialized imaging studies; and biopsy, when indicated.

Your doctor will attempt to rule out conditions with similar symptoms. These may include infections; inflammatory disorders (Crohn's disease, vasculitic syndromes, rheumatoid arthritis, systemic lupus, or others); certain cancers; and multiple sclerosis. Blood tests and urinalyses will likely be ordered, with studies to analyze liver function and test for a specific antibody related to Cogan's syndrome.

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

- Detailed exam by an ophthalmologist and an ear, nose, and throat specialist (ENT)
- Echocardiography—a diagnostic 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. Steroid medications are typically started early in the course of the disease 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/or immunosuppressive medications may be prescribed, including methotrexate, cyclophosphamide, cyclosporine, or azathioprine.
- If impaired hearing does not respond to medications, cochlear implants may help. These are surgically implanted electronic devices that provide a sense of sound to those who are deaf or hard of hearing. Medications may 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 are 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 vasculitis are proven to be present, treatments include steroids and immunosuppressive therapy. Heart problems may require surgical procedures, such as aortic valve replacement.

## COGAN'S SYNDROME

### Side effects

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 a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

### 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 lab 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, you will likely need to see the following specialists: ophthalmologist (eyes); otolaryngologist (ear, nose and throat); rheumatologist (joints, muscles, 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 with 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 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.

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

## **COGAN'S SYNDROME**

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](http://www.vasculitisfoundation.org/research)

### **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](http://www.vasculitisfoundation.org)

### **Connect with the VF on Social Media**

- Instagram: [vasculitisfoundation](https://www.instagram.com/vasculitisfoundation)
- Twitter: [@VasculitisFound](https://twitter.com/VasculitisFound)
- VF Facebook Discussion Group: [www.facebook.com/groups/vasculitisfoundation](https://www.facebook.com/groups/vasculitisfoundation)

## **COGAN'S SYNDROME**

### **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](http://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](http://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](http://www.VasculitisFoundation.org)  
Email: [vf@vasculitisfoundation.org](mailto:vf@vasculitisfoundation.org)

The VF gratefully acknowledges Alexandra Villa-Forte, MD, MPH, Cleveland Clinic, for her expertise and contribution to this brochure. This brochure was supported by an unrestricted education grant from Genentech. The Vasculitis Foundation is solely responsible for all content.

The Vasculitis Foundation (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. VF does not endorse any medications, products or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

*(Updated October 2018)*