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VASCULITIS FOUNDATION

What is Cogan's Syndrome?

Cogan's syndrome is a rare form of vasculitis that affects the eyes and inner ears, and primarily the large blood vessels—especially the aorta, which carries blood from the heart to the rest of the body.

Vasculitis is a group of rare disorders characterized by inflammation of the blood vessels. Inflammation of blood vessels may result in aneurysms or narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

Causes

The exact cause of Cogan's syndrome is not fully understood. 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 may develop 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. The disease can also occur in people of any race, but is more common in Caucasians. It is not known to run in families.

Symptoms

Cogan's syndrome typically starts with either eye inflammation or inner ear inflammation, but often progresses to include both. 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 can be present at the beginning of Cogan's syndrome or may develop later in the course of the syndrome. They include:

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- Muscle pain and cramping
- Joint pain
- Headache
- Fever
- Weight loss
- Heart murmurs or other heart problems may develop

In some cases, symptoms come and go.

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
- A physical examination exam by an ophthalmologist and an ear, nose, and throat (ENT) specialist
- Laboratory tests
- Echocardiography – a diagnostic ultrasound of the heart
- Magnetic resonance angiography to look at the 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, such as prednisone, are typically started early in the course of the disease for eye inflammation and decreased hearing.

- Mild eye disease may be treated with steroid drops in the eye. Nonsteroidal anti-inflammatory drugs may also be used.

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- 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.

Early diagnosis and treatment are important to lower the risks of permanent hearing or vision loss.

Side Effects of Treatment

The medications used to treat Cogan's syndrome have potential serious side effects, such as:

- Lowering your body's ability to fight infection
- Bone loss (osteoporosis), among others

Therefore, it is 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:

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- Ophthalmologist (eyes)
- Otolaryngologist (ear, nose, 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 and 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 is 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. This can affect your 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

There is no cure for Cogan's syndrome. Most patients respond well to treatment; however, in some cases, it can lead to permanent hearing or vision loss and chronic symptoms of vertigo. 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.

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