Churg Strauss Syndrome

What is an Eosinophil?

Eosinophils, a type of white blood cell, are an important part of the immune system, helping us fight off certain types of infections, such as parasites. Many different problems can cause high numbers of eosinophils in the blood including allergies, parasitic infection, eosinophil associated gastrointestinal disorders, leukemia, and other problems. When eosinophils occur in higher than normal numbers in the body, without a known cause, an eosinophilic disorder may be present. Eosinophilic disorders are further defined by the area affected. For instance, eosinophilic esophagitis means abnormal numbers of eosinophils in the esophagus.

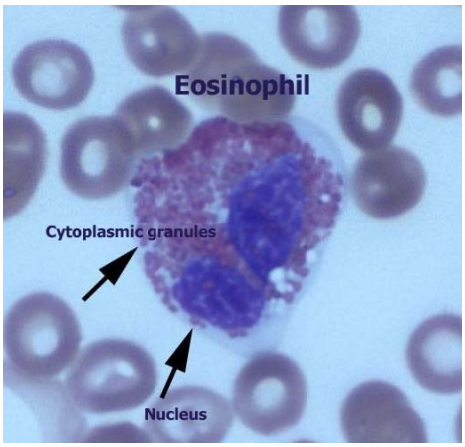

Image: *Eosinophil, Courtesy of Dr. Margaret Collins*

What is Churg Strauss Syndrome?

Churg Strauss Syndrome (CSS) is a rare systemic disease characterized by asthma, high levels of eosinophils, and inflammation of small to medium sized blood vessels (vasculitis). Eosinophils are elevated in the blood and tissues in CSS. The inflammation can affect various organ systems including the lungs, gastrointestinal tract, skin, heart and nervous system. CSS is also sometimes referred to as allergic granulomatosis or allergic angiitis. The cause is unknown and individuals may be affected very differently by CSS. With early diagnosis and treatment, CSS can be successfully managed.

Symptoms
Symptoms range from mild to a wide variety of problems depending on the organ system involved and the severity of the disease.

- Asthma
- Recurrent sinus infection
- Pneumonia
- Numbness, tingling and/or pain in the feet or hands
- Difficulty breathing
- Chronic cough
- Rashes
- Fevers
- Night sweats
- Muscle aches
- Enlarged lymph nodes
- Weakness
- Severe abdominal pain
- Gastrointestinal bleeding
- Heart problems
- Chest pain

CSS is a progressive disease consisting of three phases:

1. Prodromal (Allergic)
2. Hypereosinophilic
3. Vasculitis

The first stage, **allergic phase**, typically consists of asthma, sinusitis, allergic rhinitis, and/or recurrent respiratory infections. During this phase, the severity of problems often increases, with the asthma, sinusitis, and rhinitis becoming more difficult to treat and control.

During the next phase, the **hypereosinophilic phase,** patients may develop chronic eosinophilic pneumonia and eosinophilic gastroenteritis (inflammation of the digestive tract). Symptoms depend on organ involvement, but may include weight loss, fever, night sweats, cough or abdominal pain. Symptoms may improve and then recur over a period of months to years. With treatment, some never experience the third phase.

The third phase is the **systemic vasculitis phase** during which inflammation of blood vessels throughout the body causes damage to different organs. A biopsy is helpful in diagnosing this vasculitis, but not always necessary. Because CSS can affect many different organs at this stage, symptoms vary widely depending on the organ affected. Common organs affected by CSS include the skin, heart, lungs, nervous system, kidneys, musculoskeletal system and the gastrointestinal tract.

Diagnosis of CSS may be suspected in individuals with asthma**,** eosinophilia > 10% of white blood cell count, sinusitis, pulmonary infiltrates (hazy shadows on chest x-ray), and neuropathy. Biopsies showing vasculitis and extra vascular eosinophils make a definitive diagnosis. Anti-neutrophil cytoplasmic antibodies (ANCA) may be found in many patients. While a combination of these findings are suggestive of CSS, these findings can also be seen in other disorders including Wegener's granulomatosis, polyarteritis nodosa and eosinophilic pneumonia. Some patients with eosinophilic gastroenteritis may have high blood levels of eosinophils and associated allergic diseases, such as asthma, but do not have CSS.

Treatment

There is no cure, but many people achieve long-term remissions with medication. Treatment consists of reducing inflammation of the blood vessels and suppressing the immune system. Recent advances in diagnosis and treatment have greatly improved long-term outcomes.

Medications

1. **Prednisone**

Systemic steroids (oral or intravenous) are usually the initial therapy to reduce inflammation in CSS. Initially, high doses of oral steroids are given. The steroids are then slowly tapered down to a lower dose for maintenance. Prednisone is the most common and effective medication used to treat CSS.

2. **Immunosuppressant medications**

Cyclophosphamide (Cytoxan) is a very powerful medication that works by suppressing rapidly dividing cells in the immune system. Azathioprine (Imuran), Cyclosporine and/or Mycophenolate mofetil are also used when the disease does not respond to steroids alone. Like cyclophosphamide, they too can cause serious side effects, but the risk is lower. Regular blood tests are important to monitor dosing and potential side effects.

3. **Interferon alpha**

Interferon alpha, given by intramuscular injection or under the skin, is one of the biological drugs used in the treatment of a variety of different diseases, including leukemia. Interferon is becoming more widely used because it usually has less toxic side effects but still requires monitoring.

4. **Other** drugs used in the treatment of Churg Strauss Syndrome are not listed here. More information on the medications used to treat CSS can be found on the CSSA and APFED websites.

The initial diagnosis of CSS can be overwhelming and often affects the entire family. Early diagnosis and medical treatment can control the symptoms in most people. A positive attitude, a good support system, and proper medical care are important in learning to live with Churg Strauss Syndrome.

Content developed in conjunction with the Churg Strauss Syndrome Association.

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About APFED

American Partnership for Eosinophilic Disorders (APFED) is a non-profit organization dedicated to patients and their families coping with eosinophilic disorders. Our mission is **Education**, **Awareness**, **Support** and **Research.** All medical information is reviewed for accuracy by our medical advisory board. www.apfed.org

About the Churg Strauss Syndrome Association

The Churg Strauss Syndrome Association is dedicated to the identification, treatment and cure of Churg Strauss Syndrome. For more information, visit www.cssassociation.org

Dr. Michael Wechsler welcomes questions about Churg Strauss Syndrome. Please feel free to contact him at Brigham and Women's Hospital 75 Francis Street, Boston, MA 02115, call 617-732-8202 or by email mwechsler@rics.bwh.harvard.edu