

Churg Strauss syndrome (CSS), also called allergic granulomatosis and angiitis, is a disorder characterized by allergic rhinitis (inflammation of the nose), asthma, and an alteration in blood counts called eosinophilia. The organs involved are usually the lung and the skin although any organ system can be affected including the cardiovascular (heart), gastrointestinal (stomach), kidneys and the central nervous system.

Both men and women are equally diagnosed with CSS. The mean age at diagnosis is 50 years, but the systemic vasculitic phase is frequently apparent in patients who are in their late 30s. It is uncommon in people older than 65 years.

The disease is most likely due to an autoimmune process, which is a disorder of the immune system.

The **clinical manifestations** of CSS usually has three stages.

1. The **first phase** is called the “allergic” phase and is characterized by allergic inflammation of the nose, the skin and the lungs. People are often diagnosed with late onset asthma during this phase.
2. The **second phase** is called the “hypereosinophilic” phase, which means that there are too many eosinophils (a type of white blood cell) in the body. This phase is characterized by inflammation of the esophagus, stomach or intestines.
3. The **third phase** is the “systemic vasculitis” phase. During this phase there is inflammation and damage of blood vessels. Blood vessels can be damaged in different parts of the body. During this phase people may suffer from fever, weight loss, and lack of energy.

Asthma is the main feature of CSS (occurring in more than 95 % of patients) and usually precedes the vasculitic phase by approximately 8 to 10 years. It is frequently chronic and of sufficient severity to require long-term corticosteroid therapy.

Allergic rhinitis and skin involvement are also common. Manifestations of cardiac involvement in CSS include pericarditis, a condition characterized by inflammation of the external part of the heart, heart failure and myocardial infarction (heart attack). Dysfunction of the peripheral nerves, usually called mononeuritis multiplex, is seen in up to 75 % of patients with CSS.

Kidney involvement may be more common than is generally reported. A gastroenteritis (inflammation of the gastrointestinal tract) characterized by abdominal pain, diarrhea, bleeding, may precede or coincide with the vasculitic phase of CSS.

There are no specific **laboratory** tests for CSS. Eosinophilia, the increased percent of eosinophils, a type of white blood cells, above the normal range in blood counts is the most characteristic finding.

Antineutrophil cytoplasmic antibodies (ANCA) are found in several systemic vasculitides including CSS, and 38 to 59 % of CSS patients are ANCA positive.

The **diagnosis** of CSS is suggested by the clinical findings and then confirmed by lung biopsy or biopsy of affected tissues.

Treatment: Most patients with CSS respond favorably to corticosteroid therapy. Late relapses after a successful response to treatment are uncommon so treatment can be discontinued in most patients. Additional treatment options include inhaled steroids, cyclophosphamide (Cytoxan), azathioprine (Imuran) , and high-dose intravenous immune globulin (IVIG) have been used in patients with severe disease or disease that is unresponsive to corticosteroids. Such patients have been improved with a regimen of corticosteroids and interferon-alpha. Plasma exchange occasionally has been used in conjunction with other therapies.

All the immunosuppressive drugs include the possibility of development of infections since the immune system is weakened. In addition adverse effects of corticosteroids may include glucose intolerance or frank diabetes mellitus (condition of abnormal glucose levels in blood). Patients should be aware and watchful in order to report symptoms suggestive of diabetes or infection. Use of glucocorticoids may also predispose to, or worsen preexisting, osteoporosis, a disorder of the bone density. This is a particular concern especially for postmenopausal women and older men. Use of calcium supplements, vitamin D, hormone replacement therapy and/or bisphosphonates can help protect against bone mineral loss.

Autoimmunity is an unpredictable disorder developing under unspecified yet conditions. Psychological, environmental, genetic factors interfere and in combination with the treatment option selected lead to an individualized course and prognosis for each patient.