

EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA)

(Formerly known as Churg Strauss Syndrome)

WHAT IS EGPA?

Eosinophilic Granulomatosis and Polyangiitis (EGPA), previously called Churg-Strauss syndrome, is a disorder characterized by allergic rhinitis (inflammation of the nose), asthma, and an increase in certain blood cells known as eosinophils, which infiltrate and inflame different organ systems. The organs most commonly involved are the lungs, peripheral nerves, sinuses, and the skin, although any organ system can be affected including the cardiovascular (heart), gastrointestinal (stomach), kidneys and the central nervous system.

Asthma is the most common sign of EGPA. The disorder can also cause a variety of other problems, such as hay fever, rash, gastrointestinal bleeding, and pain and numbness in your hands and feet. Skin involvement with rashes and nodules are also common. Manifestations of cardiac involvement in EGPA includes pericarditis, a condition characterized by inflammation of the external part of the heart, heart failure and myocardial infarction (heart attack).

SYMPTOMS

This is usually the first stage of EGPA.

It's marked by a number of allergic reactions, including:

1. The first phase is called the "allergic" phase and is characterized by allergic inflammation of the nose, the sinuses, the skin and the lungs. People are often diagnosed with late onset asthma during this phase. Others may have childhood onset asthma and allergies that suddenly worsen later in life.
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 blood and in certain organs. This phase is characterized by eosinophilic inflammation of the lungs, 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, leading to organ damage from restricted blood flow. During this phase people may suffer from fever, weight loss, and lack of energy.

EGPA

CAUSES

The exact cause of EGPA is unknown. It's likely that an overactive immune system response is triggered by a combination of genes and environmental factors, such as allergens or certain medications. Instead of simply protecting against invading organisms such as bacteria and viruses, the immune system overreacts and targets healthy tissue, causing widespread inflammation.

TREATMENT

No cure for EGPA exists. But certain medications may help manage your symptoms. Most patients respond favourably to corticosteroid therapy. Many patients do relapse over time, and may require indefinite steroid therapy. If a patient has difficulty being maintained on low dose steroids alone, it is usually preferable to add another immunosuppressive drug to the regime to allow the steroids to be tapered to as low a dose as possible. 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. Plasma exchange occasionally has been used in conjunction with other therapies.

COPING AND SUPPORT

EGPA is a serious disease. Even when it's in remission, you may worry about the possibility of recurrence or about long-term damage to your heart, lungs and nerves.