Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss) Factsheet

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Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a form of vasculitis that affects the small to medium sized blood vessels which supply many organs including the heart, lungs, nerves and skin. Patients with EGPA commonly have abnormal levels of eosinophils, a type of white blood cell. It is these eosinophils which are thought to cause inflammation and consequent damage to the blood vessel wall.

Who gets EGPA?

EGPA is a rare illness. About 6 per 1000,000 adults develop the condition every year. It is slightly more common in men than in women. It usually develops between the ages of 15-70, but has been reported in patients as young as 9.

What causes EGPA?

The cause is unknown although genetic disorders and problems with the immune system have been suggested. Medicines have also been associated with the development of EGPA, especially in a group of medicines used to treat asthma called leukotriene receptor antagonists though this link has not been proven.

What are the symptoms of EGPA?

EGPA can affect different parts of the body but asthma is usually the first problem to develop. Other symptoms may come on at the same time or develop several years later. They may include a persistently runny nose, blockage of the sinuses with pain in the face, pneumonia or coughing up blood. Nodules can develop on the skin or you may notice little bruises or hives. Many other problems can occur, including disturbance of the digestive system, heart failure, kidney failure, high blood pressure, numbness and pins and needles.

How is EGPA diagnosed?

Blood screening tests frequently show a high level of a certain type of white cells called eosinophils. A blood test for ANCAs is positive in about 4 out of 10 people with EGPA. Tests for kidney function , inflammation and a urine test is also routinely undertaken.

Other tests you may be offered include chest x-rays and scans of various parts of the body.

What is the treatment for EGPA?

High doses of a steroid such as prednisolone help control CSS, and are often prescribed in combination with a group of medicines called immunosuppressants. These act by reducing the activity of the immune system. Examples of commonly used immunosuppresants include cyclophosphamide, methotrexate and mycophenolate mofetil. Azathioprine is a commonly prescribed drug when the disease has been controlled and used to reduce the chance of the disease recurrence.

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Does EGPA get better?

Prednisolone and immunosuppresants are usually effective in controlling most of the symptoms of EGPA although they can come back from time to time. Asthma is less easy to control and may persist. If complications such as heart or kidney disease develop this can result in long-term ill-health.

Further reading

Farid-Moayer M , Churg-Strauss Syndrome, Medscape, 2011

http://emedicine.medscape.com/article/333492-overview

Vaglio A, Moosig F, Zwerina J; Churg-Strauss syndrome: update on pathophysiology and treatment. Curr Opin Rheumatol. 2012 Jan;24(1):24-30. [abstract]

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