Eosinophilic Granulomatosis With Polyangiitis & You

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your rheumatology resource

What is it?

Eosinophilic granulomatosis with polyangiitis (EGPA) is a serious disease that causes inflammation of the small blood vessels. People who have EGPA have an abnormally high number of white blood cells called eosinophils. Everyone who has EGPA also has asthma.

The cause of EGPA is unknown at this time, but it is thought to be an autoimmune disease. This means that something triggers your immune system to go awry. It begins to attack small blood vessels and causes inflammation. This can cut off blood supply to tissues in the lungs, sinuses, skin, nerves and other vital organs.

EGPA is like a fire in your small blood vessels and organs

The word inflammation comes from the Latin word *inflammare* which means to light on fire. You can think of EGPA like a fire in your small blood vessels and organs. Putting out the fire of EGPA early is important. You want to get that fire out as quickly as possible so it doesn't cause damage. Once the damage from EGPA is done it cannot be reversed.

Treating EGPA aggressively is also essential. Using the same fire analogy, we've called in the fire department. Now we need to make sure we have the right tools to put out the fire. We don't want a bucket and water. We want a fire truck with a big hose. The faster we can get that fire out the less damage is done and the better things will be in the long run.

What is it going to do to me?

Most people have asthma for many years before they get EGPA. Prior to the onset of EGPA, their asthma usually gets worse. As the disease progresses, the number of eosinophils in the blood starts to rise. People may start feeling unwell and notice fatigue, achiness, low-grade fever, loss of appetite, weight loss, skin rashes or nodules, diarrhea or swollen lymph nodes.

Other symptoms of EGPA really depend on which organs are affected. If the lungs are involved, you may notice shortness of breath, a cough, or chest pain/discomfort. If the kidneys are involved, symptoms could include feeling tired, swelling of the legs or shortness of breath. The stomach and heart can also be involved, but this is rare.

Signs of EGPA can also appear on the skin as little red dots called purpura (this may look like bruises), in the nerves (sudden loss of strength) and joints (pain & swelling).

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RheumInfo.com is a free educational website where you can learn more about EGPA and treatments for the disease. The website is operated by Dr. Andy Thompson, a rheumatologist.

What can I do about it?

EGPA is a very serious disease. While there is no cure for EGPA, many people respond well to treatment. EGPA is a disease that tends to flare. That means that you may have periods where your symptoms are well controlled and other times they flare up again.

If you have EGPA, your family doctor should refer you to a specialist. A rheumatologist is one of the best people to help you manage your condition.

People with EGPA can lead active and productive lives with the right kinds of treatment. It is essential to treat EGPA early and aggressively. It's also important to keep your asthma under control with standard asthma therapies.

The primary therapy for EGPA is prednisone. This medicine is very effective at controlling the inflammation of EGPA. If your vital organs are affected, another medication called cyclophosphamide may be required. It is often used with prednisone to get the disease under control. After 6 months, cyclophosphamide is usually switched to azathioprine (Imuran) or methotrexate. In milder cases of EGPA, prednisone along with azathioprine may be all that is needed.

Here are some other recommendations on what you should do:

- **Learn as much as you can about the disease**
- **Attend your rheumatologist appointments regularly**
- **Get your blood tests regularly as suggested by your doctor**
- **Learn about the medications used to treat EGPA**

Flesch-Kincaid Reading Grade: 7.1