EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA)

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What is EGPA?

EGPA is an immune mediated disease characterized by inflammation of the small blood vessels in the body. The disease can cause the clustering of inflammatory cells in tissues, such as the lungs, to form **granulomas**, hence the term "granulomatosis".

In EGPA, there is an excess of **eosinophils** in the blood and affected tissues. Eosinophils are white blood cells that are normally produced in response to allergies or parasitic infections, however in EGPA they can contribute to widespread inflammation.

EGPA is also known as an "**ANCA-associated vasculitis**" due to the presence of ANCA antibodies in the blood in around 40% of the patients (only). There are 2 types of ANCAs – p/MPO-ANCA and c/PR3-ANCA. EGPA is primarily associated with MPO-ANCA.

Who is affected?

EGPA is a rare condition affecting 10-25 per million people. Men and women are affected equally. The average age of diagnosis is around 50 years.

Which organs are affected?

The lungs, nostrils and sinuses, heart, skin and nerves are most commonly affected. Essentially all individuals have adult-onset asthma.

What are the symptoms?

Depending on which organ systems are affected, symptoms and findings may include:

- Nasal congestion and/or polyps
- Shortness of breath, cough
- Wheezing and asthma attacks
- Chest pain, palpitations
- Skin rash resembling small bleeding spots, or hives and urticarial lesions that appear most often on the arms, feet and/or legs
- Burning pain, numbness or tingling of the extremities
- Weakness or inability to move the wrist or ankle, otherwise known as a "wrist drop" or "ankle drop"
- Fevers, weight loss & unusually severe fatigue

Individuals often experience an **initial period of allergic symptoms and asthma**, before developing inflammation in other systems. This initial period can last several years!

How is EGPA diagnosed?

Diagnosis is made based on a combination of the following:

1. Compatible symptoms
2. Abnormal labs, including:
   a. High inflammatory markers (ESR and/or CRP)
   b. High eosinophil count
   c. Detection of ANCA antibodies in blood
   d. High troponin and creatine kinase (CK) if the heart is affected
3. Pulmonary function testing to confirm asthma
4. Examination of the nasal passage to look for polyps
5. Imaging of the sinuses, lungs, heart and/or other affected sites.
6. Supportive features on biopsy of affected organ(s). Common sites of biopsy include the nasal passage, skin or nerves.

Remember that any of the listed symptoms by itself can be seen in more common conditions such as infections, chronic lung disease and cancers. It is therefore important to consider the "whole picture" and rule out other more common conditions before making the final diagnosis of EGPA.

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How is EGPA treated?

The general treatment protocol consists of 2 phases:

1. **Induction phase** – This phase is aimed at rapidly suppressing the inflammation and "inducing" the disease into a state of dormancy (ie. "remission"). High dose prednisone with gradual tapering is started alongside another immunosuppressive medication. Cyclophosphamide may be used in severe cases.
2. **Maintenance phase** – At this point, the disease is inactive and the goal is to maintain remission. Common medications used for maintenance include methotrexate, azathioprine and mycophenolate mofetil.

Adjunctive therapies include:

- **Asthma** therapy is crucial and includes puffers or special injection therapies. Your asthma specialist will determine if you are eligible for these.
- **Pneumocystis pneumonia (PCP) protection** with an antibiotic when taking cyclophosphamide.
- **Bone and stomach protectants** while on high dose prednisone.
- **Nasal rinses & sprays** for nasal and sinus symptoms.

How is my EGPA monitored?

Your rheumatologist often works with a **respirologist** and **allergy doctor** to treat your EGPA. Depending on which organs are affected, other specialists may be involved.

Your **family physician** also plays an important role in monitoring for high blood pressure & cholesterol, diabetes and osteoporosis.

Common tests used to monitor the disease include:

- Lab tests for blood counts (including eosinophil level), inflammatory markers, liver & kidney function, ANCA levels
- Urine studies for blood & protein
- X-rays and CT scans
- Echocardiograms
- Pulmonary function tests
- EMG (nerve studies)
- Nasal scopes (if needed)

Testing is typically more frequent at the beginning or when there is active disease, and less frequent once the disease is stable and in remission. Biopsies are typically done for diagnostic purposes and do not need to be repeated unless there is a new or worsening feature that needs to be defined further.

Is EGPA a fatal disease?

The survival rate for EGPA is high when treated. The outcomes depend on a variety of factors, including the severity of disease, whether major organs are affected (eg. heart, intestines or kidney), delays in diagnosis and treatment, and complications (eg. infections) during treatment.

What will happen to me? How long do I need to stay on medications?

Like other autoimmune diseases, EGPA is treatable but not yet curable. Symptoms and disease activity resolve with medication, however may return when the medication is reduced or stopped. When the disease returns ("flares"), it is called a **relapse**.

Currently, we do not know the optimal duration of immunosuppressive treatment for EGPA. Up to 40-50% of patients may relapse within the 1st year, and sometimes this is caused by stopping the prednisone or reducing the dose.

The asthma component of the disease is often the hardest to treat and control, and most patients require lifelong asthma therapy.

EGPA DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Nasal congestion and polyps
- ☐ Sinus congestion
- ☐ Skin rash and/or ulcers
- ☐ Large painful hives lasting over 24 hours
- ☐ Shortness of breath
- ☐ Coughing or asthma attacks
- ☐ Chest pain or palpitations
- ☐ Burning, tingling or numbness of the hands or feet
- ☐ Weakness of the arm or leg
- ☐ Joint pain or joint swelling
- ☐ Abdominal pain, especially after eating
- ☐ Bloody stools
- ☐ Fevers or night sweats
- ☐ Weight loss
- ☐ Unusually severe fatigue
- ☐ Others: _____

Medications that have been prescribed:

	Date started	Date stopped
IV methylprednisolone _____ mg		
Prednisone: (Starting dose _____ mg)		
Methotrexate ____ mg		
Folic acid (only if taking methotrexate)		
Mycophenolate mofetil _____ mg		
Azathioprine ____ mg		
Leflunomide ____ mg		
Cyclophosphamide _____ mg		
Septra/Bactrim/Sulfatrim		
Mepolizumab _____ mg		
Omalizumab _____ mg		
Benralizumab _____ mg		
Rituximab _____ mg		
Others:		

Apart from lab tests, I have had the following investigations:

- ☐ Echocardiogram
- ☐ Bronchoscopy
- ☐ Nasolaryngoscopy (nasal scope)
- ☐ Pulmonary function test
- ☐ Electromyography (EMG/nerve conduction study)
- ☐ X-rays (circle all that apply): Chest | Abdomen | Joints
- ☐ CT scans (circle all that apply): Chest | Abdomen | Head | Sinuses
- ☐ MRI (circle all that apply): Brain | Heart | Abdomen
- ☐ Biopsy (circle all that apply): Skin | Nasal polyp | Kidney | Lung Nerve | Bone marrow

What do I need to do?

- ✓ Attend **regular** follow-up visits with my rheumatologist and other specialists.
- ✓ **Do regular** blood and urine tests for disease and medication monitoring.
- ✓ Take my **asthma medications** regularly and avoid usual triggers for my asthma attacks
- ✓ Ensure my medical team is monitoring me for diabetes, high blood pressure and high cholesterol.