GRANULOMATOSIS WITH POLYANGIITIS (GPA)

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What is GPA?

GPA is a type of vasculitis affecting the smallest blood vessels in the body. The disease can cause the clustering of inflammatory cells in tissues, such as the lungs or sinuses, to form **granulomas**, hence the term "granulomatosis".

GPA is also known as an "ANCA-associated vasculitis", due to the presence of ANCA antibodies in the blood. There are 2 types of ANCAs – p/MPO-ANCA and c/PR3-ANCA. GPA is associated more often with PR3-ANCA.

Which organs are affected?

GPA can cause inflammation and granuloma formation in essentially all organs. The sinuses, nasal cavity, respiratory tract, kidneys, and skin are particularly affected.

Who is affected?

GPA is a rare condition affecting 50-200 per million people in Western countries. It is more common in people of European descent. Men and women are affected equally. Most develop this condition between the ages of 40 to 65 years . Cases in children and young adults are rare.

What are the symptoms?

GPA can cause a variety of symptoms depending on which organs are affected. Possible symptoms include:

- Sinus congestion & sinusitis
- Nasal pain, crusting & bleeding
- Changes in the shape of the nose due to cartilage destruction (saddle nose deformity, nasal septum perforation)
- Shortness of breath, cough and bloody sputum
- Recurrent ear infections
- Hearing loss, or a "whooshing" / ringing sound in the ears
- Painful red (bloodshot) eye
- Joint pain & swelling
- Skin rash resembling small bleeding spots on the feet and legs
- Unusually bubbly / frothy / dark urine
- Fevers, weight loss & unusually severe fatigue

Many of the above symptoms can also be seen in infections, chronic lung diseases and cancers. It is therefore important to consider the "whole picture" and rule out other possible diagnoses. Symptoms can present very suddenly over a short period of time or can be staggered over years. This can make it difficult to timely diagnose the disease.

How is GPA 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 ANCA (mostly c/PR3-ANCA
   c. High creatinine with protein & blood in the urine (if kidneys are involved)
3. Imaging of the lungs and/or other affected sites.
4. Bronchoscopy and/or nasal scope to assess for inflammation in the airways.
5. Supportive features on biopsy of affected organ(s), such as the kidneys, nasal cavity, lungs or skin. Biopsy may not be needed if there is enough defining features based on symptoms and other results.

Are there different forms of GPA?

GPA can be classified as "**Limited**" – ie. mainly localized to the ears, nose and throat tissues; or "**Systemic**" where any organ can be affected.

ANCA can be negative in limited GPA but is almost always positive in systemic GPA. The two forms also differ with respect to treatment and prognosis.

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How is GPA treated?

There are 2 phases of treatment:

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 is started alongside another immune suppressing medication. If your disease is severe, cyclophosphamide or rituximab may be used. The prednisone dose will be lowered over several months' time.
2. **Maintenance phase** – At this point, the disease is inactive and the goal is to maintain remission. Common maintenance medications include rituximab, azathioprine, methotrexate, and mycophenolate mofetil.

Adjunctive therapies include:

- **Pneumocystis pneumonia (PCP) protection** with an antibiotic if on rituximab or cyclophosphamide.
- **Nasal rinses & sprays** for nasal and sinus symptoms.
- **Dialysis** if there is severe kidney disease.
- **Plasma exchange** may be used in refractory, life-threatening cases (although no clear benefits were seen in recent studies).

How is my GPA monitored?

Your rheumatologist often works with other specialists to treat your GPA. Depending on which organs are affected by the disease, you may expect to also see a **respirologist, nephrologist** or **otolaryngologist**, among others.

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, inflammatory markers, liver & kidney function and ANCA levels
- Urine studies for blood and protein
- X-rays, CTs and ultrasounds
- Pulmonary function tests (breathing tests)
- Nasal scopes

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.

The ANCA level often normalizes once the disease is in remission and can increase during a flare. However, up to 40% of patients have a persistently elevated ANCA level despite remaining in long-term remission.

Is GPA a fatal disease?

The survival rate for GPA is high when treated – up to 90% at 1 year, and up to 80% at 5 years. Outcomes depend on a variety of factors, including severity of disease, which organs are affected, delays in diagnosis and treatment, and complications (eg. infections) encountered during treatment.

What will happen to me? How long do I take medications for?

Like many autoimmune diseases, GPA is treatable but not yet curable. Symptoms resolve with medications, however can return when the medication(s) are reduced or stopped. When the disease returns ("flares"), it is called a **relapse**.

Currently, we do not know the ideal duration of maintenance treatment for GPA. 1 in 5 patients (20%) will relapse within the 1st year of diagnosis, and up to 3 in 5 patients (60%) will relapse within 5 years. Because of this, most patients are kept on medications for at least 2-5 years.

Patients who have organ damage or have had multiple relapses already may be kept on treatment longer (or even lifelong).

GPA DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Nasal congestion, crusting or bleeding
- ☐ Sinus congestion and sinusitis
- ☐ Ear infections
- ☐ Hearing loss, sensation of ear fullness
- ☐ Protrusion of the eyes
- ☐ Painful, red (bloodshot) eyes
- ☐ Hoarse voice
- ☐ Shortness of breath
- ☐ Cough and/or coughing up blood
- ☐ Skin rash and/or skin ulcers
- ☐ Burning, tingling or numbness of the hands or feet
- ☐ Weakness of the arm or leg
- ☐ Joint pain or joint swelling
- ☐ Unusually bubbly / frothy or dark urine
- ☐ Fevers or new night sweats
- ☐ Weight loss
- ☐ Severe fatigue
- ☐ Others: _____

Medications that have been prescribed:

	Date started	Date stopped
IV methylprednisolone _____ mg		
Prednisone: (Starting dose _____ mg)		
Cyclophosphamide _____ mg		
Rituximab _____ mg (INDUCTION)		
Rituximab _____ mg (MAINTENANCE) (Number of infusions: _____)		
Septra/Bactrim/Sulfatrim		
Methotrexate ____ mg		
Folic acid (only if taking methotrexate)		
Mycophenolate mofetil _____ mg		
Azathioprine ____ mg		
Leflunomide ____ mg		
Plasma exchange (PLEX) (Number of sessions: _____)		
Others:		

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

- ☐ Scopes (circle all that apply): Bronchoscopy | Nasal scope
- ☐ Audiology test
- ☐ 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 | Spine | Orbits (eyes)
- ☐ Biopsy (circle all that apply): Skin | Nasal | Kidney | Lung Trachea | Bronchus | Other: _____

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.
- ✓ Ensure my medical team is monitoring me for diabetes, high blood pressure and high cholesterol.
- ✓ Seek urgent medical attention if you are having daily fevers, coughing up blood or experiencing other concerning symptoms.