# BASELINE TESTS

| IMPORTANT BASELINE TESTS FOR ALL VASCULITIS | OPTIONAL: BASED ON CLINICAL PRESENTATION |
|---|---|
| • **ANCA blood panel** (ELISA): to idenitify if a person is ANCA positive. ANCA negative does not necessarily exclude the possibility of vasculitis<br>  ◦ MPO (myeloperoxidase) – common in MPA, occasionally in EGPA<br>  ◦ PR3 (proteinase-3) – common in GPA<br>• **Complete Blood Count (CBC) with Differential**– identify infection and elevated eosinophils (EGPA)<br>• **Comprehensive Metabolic Panel** – evaluate kidney and liver function, electrolytes and other key levels<br>• **Troponin, CK** – to detect prior heart attack<br>• **Inflammation markers: ESR** (sedimentation rate), **CRP** (C-reactive protein),<br>• **C3, C4** - systemic test to identify level of inflammation in the body.<br>• **Urinalysis** - evaluate kidney health<br>• **Chest X-ray** – detect infiltrates, pulmonary edema, nodules, effusions<br>• **CT Chest** - looks for lung damage and eosinophilic infiltrates (if chest xray or lung function test is abnormal)<br>• **EKG** - measures electrical activity in the heart and looks for heart problems<br>• **Echocardiogram (Echo, Heart Ultrasound)** – to evaluate heart involvement, particularly in EGPA<br>• **Pulmonary Function Tests (PFT)** – critical for EGPA, assess lung involvement, obstructive or restrictive patterns | • **TB & Hepatitis Panel** – required prior to starting some immunosuppression therapeutics<br>• **ds-DNA** - can help to rule out other autoimmune diseases<br>• **Histone Ab** - helps to rule out other autoimmune diseases<br>• **CTD screen Elia** - to detect autoantibodies associated with autoimmune connective tissue diseases<br>• **CT abdomen/pelvis** – to evaluate kidneys and or possible abdominal vasculitis involvement<br>• **CT Sinus** – important in EGPA and GPA to identify granulomas or inflammation, polyps and blockage<br>• **Head CT** - to look at detailed images of brain tissue<br>• **Cardiac MRI** - to see detailed images of heart and blood vessels to identify problems<br>• **Ultrasound with Renal Doppler** – assess renal perfusion, detect renal artery involvement<br>• **Biopsy** - Kidney, Lung, Skin particularly before treatment begins<br>• **DEXA Scan** (within 6 months of starting steroids)<br>  ◦ It is important to ensure patients on steroids take supplements to help prevent steroid damage in bones: calcium (1,000 mg) and vitamin D (2,000 IU) daily if prednisone is prescribed (confirm with rheumatologist)<br>• |

DECISION TREE

VASCULITIS
Best Practice Suggestions

# HELPFUL MEDICAL TERMS

## APPEAL (INSURANCE)
Your right to fight back if insurance denies coverage. You (or your provider) submit more information or explain why the treatment is medically necessary. Appeals can be successful, especially when supported with strong documentation. A good heartfelt letter from the patient perspective helps!

## BASELINE TESTS
Initial medical tests done after diagnosis (or at the start of a new care phase) to understand your current organ function and disease activity. These tests help measure progress or flare-ups over time.

## CARE TEAM
All the people involved in your care—this may include a variety of specialists, your PCP, pharmacists, therapists, and prior-authorization staff at your doctor's office, and more.

## DENIAL (INSURANCE)
When your insurance company refuses to pay for a treatment or test your doctor has recommended. This can feel defeating, but it doesn't mean the end of the road.

## DISEASE STATUS
Refers to how active your disease is right now. It can be described as managed, occasional flares/still on steroids, unmanaged. Understanding this helps you and your doctor decide what treatment or monitoring is needed.

## IN-NETWORK
A doctor, hospital, or provider that has a contract with your insurance company. You'll often pay less when with in-network providers.

## OUT-OF-NETWORK
A provider that does **not** have a contract with your insurance. This usually means **higher costs** for you, and in some cases, your insurance may not cover it at all.

## FLARE
When your disease suddenly gets worse after being stable or improving. You might feel new symptoms, old ones returning, or everything just feels "off." A flare can be physical, emotional, or medical – your lab results might show increased inflammation or disease activity.

## HEAT KITS
**Hospital Emergency Advocacy and Treatment Kits.** Customized kits designed to help patients with rare or complex diseases in emergency or hospital settings. They include disease specific information as well as a mechanism to keep all of your medical information in one convenient, succinct and easy to grab place.

## MANAGED DISEASE
Your disease is under control with treatment and without steroids. Symptoms are stable or improving, and you're able to live your life with less disruption.

## PRIOR AUTHORIZATION

A process where your doctor must get approval from your insurance before they will pay for certain medications, tests, or procedures. It can cause delays, but it's often required for costly or specialty treatments.

## QUALITY OF LIFE

How your disease and treatment affect your daily life—physically, emotionally, socially, and mentally. It's more than symptoms—it includes how well you're sleeping, working, connecting with others, and doing what matters to you.

## RELEASE OF INFORMATION

A legal process where you **give permission** for your medical records to be shared with others involved in your care. You sign a form that explains what's being shared, who it's going to, why, and for how long. ROI helps ensure coordinated care—**with your full consent**. It is important to clarify if it is for all information.

## REMISSION

This is a controversial term as there is not standard definition. It is often described as no disease activity and on a stable amount of medication as determined by physician. This does not mean the disease is cured or is gone, it is merely quiet.

## ROUTINE MONITORING

Regular labs or imaging to make sure your disease is controlled and treatment is working. It helps catch problems early and guide adjustments to your care plan. Your Quality of Life tool should be part of your routine monitoring.

## TEAM LEAD

A main point of contact on your care team who helps coordinate your vasculitis care. They are usually a rheumatologist. For a person with GPA or MPA it may be a nephrologist. For a person with EGPA it may be a pulmonologist.

## TREATMENT GUIDELINES

Evidence-based recommendations developed by medical experts that outline best practices for diagnosing, treating, and monitoring your disease. While helpful, they may not fit every patient perfectly.

## TREATMENT OPTIONS

Different choices for managing your disease. These might include medications, infusions, lifestyle changes, surgeries, or clinical trials. The treatment options will include newer medications not yet incorporated into treatment guidelines.

## UNMANAGED DISEASE

Your disease is not under control—symptoms may be worsening, flaring, or affecting your daily life significantly. This might mean your current treatment isn't working or more support is needed.

## VASCULITIS CENTER

A medical center that specializes in diagnosing and treating vasculitis. These centers have experts with experience managing complex vasculitis cases. They can act as a consultant to a local provider who sees fewer vasculitis patients.

# HEALTH CARE TEAM MEMBERS

You will not see all of these providers, you will only see the ones you need based on your symptoms and disease issues.

| Provider or Team Member | Why They Matter |
|---|---|
| **Primary Care Doctor (PCP)** | Keeps track of overall health and coordinates care with specialists. |
| **Rheumatologist** | Leads vasculitis care, especially if many body systems are involved. |
| **Nephrologist** | Manages kidney problems and can lead care if kidneys are affected. |
| **Pulmonologist** | Helps with breathing issues, asthma, or lung symptoms. May lead EGPA care. |
| **ENT Specialist** | Treats nose, ear, and sinus issues, common in GPA, MPA & EGPA. |
| **Allergist** | Helps manage allergies, asthma, and immune system issues. |
| **Cardiologist** | Checks and treats heart issues, especially if tests show heart involvement. |
| **Neurologist** | Helps with nerve problems like numbness, weakness, or foot drop. May lead GPA, MPA care. |
| **Dermatologist** | Treats rashes, ulcers, and other skin issues. |
| **Ophthalmologist** | Treats eye problems like inflammation or pain. Screen for cataracts and glaucoma due to steroids. |
| **Endocrinologist** | Helps with hormone problems like diabetes or bone health and steroid withdrawal, adrenal insufficiency. |
| **Dentist** | Checks for mouth issues from meds or disease, like infections or bone loss. |
| **Mental Health Therapist** | Emotional support, grief and life changes—especially helpful with stress or chronic illness. |
| **Pharmacist** | Explains meds, checks for bad interactions, and helps educate to ensure they are taken them safely. |
| **Prior Authorization Staff** | Gets insurance approval for meds and treatments. This is a staff member from the doctor's office. |
| **Insurance Case Manager** | Helps with insurance, appointments, and getting the care needed. This is a staff member from your insurance company. |

| TOPIC OF THE TIP | TIP |
|---|---|
| *Vasculitis Center* | **Vasculitis Centers** often offer access to the most current treatments and clinical trials. If you're considering this option, talk to your **Team Lead** about identifying a center. |
| *Travel costs to Vasculitis Center* | **Worried about travel costs?** Fundraising through **GoFundMe** or similar platforms can help. Many people want to support you, and donating gives them a way to help when they feel helpless. |
| *Physician coverage* | **Call your insurance company** to confirm the center is **in-network.** |
| *Second opinions* | Most insurance plans offer **second opinions**, and some vasculitis experts work through national second-opinion services (scan the QR code for more information). |
| *Long wait times* | **Is your wait to see your doctor longer than 1 month?** Ask your primary care provider (PCP) to help **expedite the referral.** Delays in starting treatment can cause **irreversible organ damage.** |
| *Home monitoring* | **Home monitoring tools** can be useful to help track symptoms and disease. Ask your Team Lead if you these and other items might be helpful: blood pressure cuff, pulse oximeter, thermometer. |
| *Contacting your doctor. Be an advocate for yourself.* | **Don't hesitate to contact your doctor** between appointments. Timely communication matters. Use your portal, call, text, or email. Speak up for yourself, this is your body and your life! |
| *Visible symptoms* | If you have **visible symptoms**, take photos (e.g., of your skin or eyes) and share them with your doctor. |
| *Invisible symptoms* | For symptoms that don't show up on tests or labs, **describe them as specifically as possible.** This helps your doctor determine if it's active disease, irreversible damage from before treatment, or something that might respond to supportive care or symptom-targeted treatment. Remember prednisone can cause weight gain, mood fluctuations, anxiety and depression, Talk with your doctor if you experience this. |
| *Be Vigilant About Your Symptoms* | You may wonder if a symptom is related. Do not dismiss new symptoms. they could be important. Talk with your doctor and inform them of any new symptom. They can help you determine if it is part of the disease. |

# MONITORING YOUR DISEASE

| Test Type | Why It Matters | How Often |
|---|---|---|
| Inflammation Markers & Infection Risk | Check if inflammation is active (CRP, ESR), IgG3, IgG4 | Every 1–3 months (more if flaring) |
| ANCA Titers | Help track disease but aren't always reliable alone (MPO, PR3). | At diagnosis & during flares |
| Renal Function | Look for kidney damage (creatinine, eGFR). | Every 1–3 months |
| Urine Tests | Check for blood or protein in urine—signs of kidney inflammation. | Every 1–3 months |
| Complete Blood Count (CBC) | Check for anemia (platelets)or blood count (white blood count, etc) abnormalities from disease or medications, | Monthly to every 3 months |
| Comprehensive Metabolic Panel (CMP) | Watch for liver (ALT, AST, ALO, Kidney (bilirubin) and other problems from medications or the disease. | Every 1–3 months |
| Pulmonary Monitoring | Look for lung issues or flare-ups (X-ray, CT, PFT/breathing tests). | Imaging yearly or as needed; PFTs at baseline or PRN |
| Cardiac Monitoring | Echocardiogram (EGPA), EKG. Screen for heart issues like inflammation. | At baseline or if symptoms show |
| Bone Health | Check for bone loss from steroids (DEXA scan). | At start and every 1–2 years |
| Eye Check | Check for eye inflammation. | At baseline and as needed |
| Vaccination Review | Stay up to date to avoid infections (Flu, Pneumonia, COVID-19, potential PJP, Shingles and RSV) | Yearly or as recommended. Discuss with rheumatologist. |