POLYARTERITIS NODOSA

What is polyarteritis nodosa (PAN)?

Polyarteritis nodosa (PAN) is a form of vasculitis—a family of rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. PAN affects medium-sized blood vessels that supply the skin, nervous system, joints, kidneys, gastrointestinal (GI) tract, and heart, among other organs. Depending on the form of the disease, PAN may affect only the skin, a single body organ, or multiple organ systems.

When PAN is systemic (affecting the whole body), symptoms can be wide-ranging, from fever, fatigue, weakness and weight loss, to muscle and joint aches, skin lesions, numbness, and abdominal pain. High blood pressure is common due to kidney damage. Individuals with PAN are also at risk for an aneurysm—an abnormal bulge in a weakened artery wall that can rupture or hemorrhage.

Prompt diagnosis and treatment are essential to prevent serious complications associated with PAN. The standard course of treatment includes corticosteroids such as prednisone used in combination with other medications that suppress the immune system. Even with effective treatment, PAN can be a chronic condition with periods of relapse and remission, so ongoing medical care is necessary.

Causes

The exact cause of PAN is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder, a disease which occurs when the body's natural defense system mistakenly attacks healthy tissue. The inflammatory process may be set in motion by a reaction to certain drugs or vaccines, or a bacterial or viral infection. PAN has been associated with hepatitis B infection. In most cases, however, the cause is unknown.

Who gets PAN?

PAN is a rare disorder, with an estimated incidence of 3 to 4.5 cases per 100,000 people annually in the United States. PAN can occur at any age, however it is more likely to affect individuals who are between the ages of 45 to 65 years. The disease is more common in men than women.

Symptoms

Because PAN can affect so many organ systems, its symptoms may be wide-ranging. At onset, patients may experience fever, night sweats, weight loss, skin sores, and severe muscle and joint pains that develop over a period of weeks or months.

Other common symptoms include:

- Fatigue
- Loss of appetite
- Abdominal pain
- High blood pressure
- Blood in the stool
- Testicular pain in men
- Chest pain
- Difficulty breathing

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- Numbness or tingling of the hands or feet
- Sudden loss of strength in the hands or feet

Complications

Potential complications can include aneurysms in the arteries that lead to the kidneys, liver or GI tract; abdominal symptoms such as pain, nausea, vomiting, or bleeding; blood clots in affected arteries; and tissue damage or loss in affected areas. The lungs are often spared.

Diagnosis

There is no single test for diagnosing PAN. Your doctor will consider a number of factors, including symptoms; a detailed medical history; physical examination; laboratory tests; and imaging studies. Your doctor will also attempt to rule out diseases with similar symptoms or characteristics. Laboratory work, including blood tests and urinalyses, can help support a diagnosis of PAN, but are not conclusive on their own. A definitive diagnosis of PAN usually requires a specialized imaging study or tissue biopsy.

Imaging: If PAN is suspected, an angiogram—an X-ray taken during the injection of a contrast agent—may help confirm diagnosis. An angiogram can detect narrowing of the blood vessels or aneurysms. Computed tomography angiogram (CTA) or magnetic resonance angiogram (MRA) scans with dye may also be used to look for changes in blood vessels.
Biopsy: A biopsy of affected tissue—surgical removal and analysis of a small tissue sample from an affected blood vessel or organ—will usually be performed to confirm the diagnosis. For PAN, most biopsies are taken from skin, nerve or muscle tissue.

Treatment

Treatment for PAN consists of corticosteroids such as prednisone to reduce inflammation. In more severe cases, prednisone may be combined with cyclophosphamide, a chemotherapy-type drug that blocks abnormal growth of certain cells, or other immunosuppressants such as methotrexate or azathioprine. High blood pressure is also treated. When PAN is related to hepatitis B, treatment often involves steroids, antiviral medications to treat the hepatitis, and sometimes plasma exchange.

Side effects

The medications used to treat PAN have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapses can occur in individuals with PAN. If initial symptoms return or new ones develop, report them to your doctor as soon as possible. Regular check-ups and ongoing monitoring of lab and imaging tests are important in detecting relapses or new organ involvement

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Your medical team

Effective treatment of PAN may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, patients with PAN may need to see the following specialists: rheumatologist (joints, muscles, immune system); cardiologist (heart); gastroenterologist (digestive system); nephrologist (kidneys); dermatologist (skin); neurologist (brain/nervous system); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

Living with PAN

Living with a chronic condition such as PAN can be overwhelming at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

There is no cure at this time for PAN, but with prompt treatment and careful monitoring, many patients can achieve remission. The long-term prognosis depends on the severity of the organs involved. Because relapses can occur, follow-up medical care is essential.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

For more information on vasculitis research, visit: www.vasculitisfoundation.org/research

VASCULITIS FOUNDATION
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About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

About the Vasculitis Foundation

The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services.

To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

Connect with the VF on Social Media

- Instagram: vasculitisfoundation
- Twitter: @VasculitisFound
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

Join the VPPRN!

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

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