

What is Polyarteritis Nodosa?

Polyarteritis nodosa (PAN) is a form of vasculitis, which is an inflammation of the blood vessels. Inflammation of blood vessels can result in aneurysms or narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

PAN affects medium-sized blood vessels that supply blood to 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.

Causes

The exact cause of PAN is not fully understood, and in most cases, the cause is unknown. It is classified as an autoimmune disorder, which is a disease that occurs when the body's natural defense system mistakenly attacks healthy tissue.

The inflammatory process may be caused by a reaction to certain drugs or vaccines, or a bacterial or viral infection. PAN has been associated with hepatitis B infection.

Who Gets PAN?

PAN is a rare disorder that can occur at any age; however, it is more likely to affect individuals who are between the ages of 45 and 65. The disease is more common in men than women.

Symptoms

Because PAN can affect so many organ systems, its symptoms may be wide-ranging and develop over a period of a few weeks to months. At the beginning, patients may have:

- Fever
- Night sweats
- Weight loss
- Skin sores
- Severe muscle and joint pains that develop over a period of a few 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
- Numbness and/or tingling of the hands or feet
- Sudden loss of strength in the hands or feet

Complications

Potential complications can include aneurysms (a weakening of the walls of the arteries, causing them to bulge). Blood clots in the affected blood vessels can cause damage to the organs affected, which can include the kidneys, GI tract, or heart. The lungs are usually not involved.

Diagnosis

There is no single test for diagnosing PAN. Your doctor will consider a number of factors, including your symptoms, a detailed medical history and physical examination, as well as:

- Laboratory work, including blood tests and urinalyses, which 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, may include:
 - Angiogram—an x-ray taken during the injection of a contrast agent into the blood vessels, which may help confirm diagnosis. An angiogram can detect narrowing of the blood vessels or aneurysms.
 - Computed tomography angiogram.
 - Magnetic resonance angiogram scans with dye may also be used to look for changes in blood vessels.
- A biopsy of affected tissue, which is the surgical removal and analysis of a small tissue sample from an affected blood vessel or organ. This will usually be performed to confirm the diagnosis, but for PAN, most biopsies are taken from skin, nerve or muscle tissue.

Your doctor will also try to rule out diseases with similar symptoms or characteristics.

Treatment

Treatment for PAN consists of:

- Corticosteroids such as prednisone to reduce inflammation.
- In more severe cases, prednisone is combined with cyclophosphamide, a chemotherapy-type drug that blocks abnormal growth of certain cells.
- Immunosuppressants such as methotrexate or azathioprine are commonly used in non-severe cases.
- High blood pressure is also treated.
- When PAN is related to hepatitis B, treatment often involves steroids and antiviral medications to treat the hepatitis.

Side Effects

The medications used to treat PAN have potentially serious side effects, such as:

- Lowering your body's ability to fight infection
- Potential bone loss (osteoporosis), among others

It is 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

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-stomach and bowels)
- Nephrologist (kidneys)
- Dermatologist (skin)
- Neurologist (brain/nervous system)
- Others as needed

The best way to manage your disease is to actively partner with your health care providers and 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.

Make a list of questions before your doctor's visit. Bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is 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. This can affect 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 (outlook) depends on the severity of the organs involved. Because relapses can occur, follow-up medical care is essential.

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