

Recommendations for managing polyarteritis nodosa (PAN)

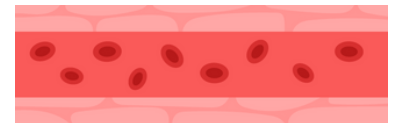


These are recommendations written by a group of doctors and patients based on research studies for people with polyarteritis nodosa (PAN).

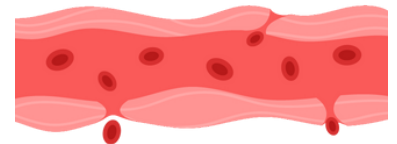
These recommendations suggest the best treatment for most people, but your individual situation and treatment may be different. Talk to your doctor about what treatment is best for you.

What is polyarteritis nodosa (PAN)?

Polyarteritis nodosa (PAN): A type of vasculitis that mostly affects medium-sized vessels, such as the vessels in your skin, nervous system, joints, kidneys, gastrointestinal (GI) tract, and heart.



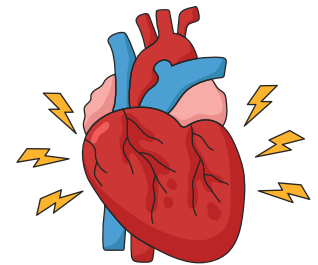
Healthy blood vessel



Inflamed blood vessel

Words to know about vasculitis

- **Vasculitis:** A group of conditions that involves inflammation (swelling) of your blood vessels.
- **Suspected disease:** Symptoms that suggest PAN and are not explained by other conditions.
- **Active disease:** New, ongoing, or worsening signs or symptoms.
- **Severe disease:** Symptoms that may cause death or organ failure (when a major organ stops working, such as the heart).
- **Nonsevere disease:** Symptoms not likely to cause death or organ failure.
- **Remission:** When symptoms go away.
- **Refractory disease:** When symptoms do not get better with treatment.
- **Relapse:** When symptoms return after a period of remission.



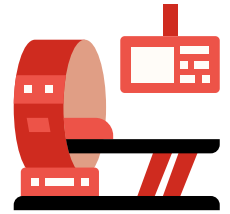
See a glossary of health terms at the end of this document.



Diagnostic recommendations

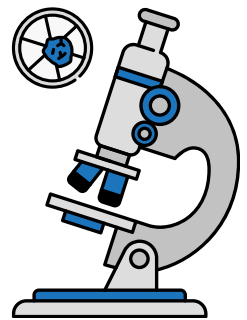
If you have **suspected PAN**:

- Use abdominal vascular imaging to help with the diagnosis and to know how serious the disease is.



If you have **suspected PAN with skin-related symptoms**:

- Get a deep or double punch skin biopsy, over a superficial (shallow) skin punch biopsy.
 - A deep or double punch biopsy can reach the medium-sized vessels in your skin affected by PAN. This can help doctors know if there is inflammation (swelling) in those vessels.



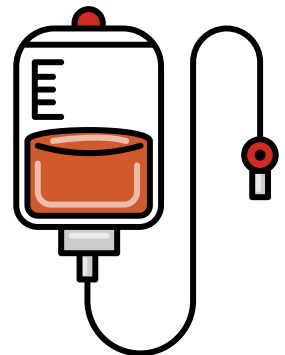
If you have **suspected PAN and peripheral neuropathy**:

- Get a nerve **and** muscle biopsy over a nerve biopsy alone.

Treatment recommendations for active disease

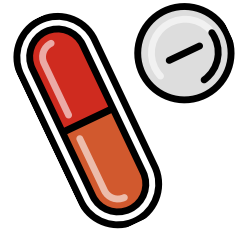
If you have **newly diagnosed active, severe PAN**, start treatment with:

- IV pulse prednisone, over high-dose oral prednisone.
 - IV prednisone may help to get severe PAN under control faster.
- Cyclophosphamide and high-dose prednisone, over high-dose prednisone alone.
 - Cyclophosphamide may help prevent disease relapse.
 - It may also lower the dose of prednisone you need, which can lower the chance of prednisone side effects.
- Cyclophosphamide and prednisone, over rituximab and prednisone, because we do not know how well rituximab works in PAN.



If you have newly diagnosed active, severe PAN but can't tolerate cyclophosphamide:

- Use other immunosuppressants (such as azathioprine or methotrexate) and prednisone, over prednisone alone.
 - Using immunosuppressants with prednisone may lower the dose of prednisone you need, which can lower the chance of prednisone side effects.



If you have newly diagnosed active, severe PAN:

- Use cyclophosphamide and prednisone alone instead of with plasma exchange, because we do not know how well plasma exchange works in PAN.
 - When **might** I get plasma exchange?
 - If your symptoms are extremely bad and have not improved despite treatment with strong immunosuppressants.



If you have newly diagnosed active, nonsevere PAN:

- Start treatment with an additional immunosuppressant (such as methotrexate or azathioprine) and prednisone, over prednisone alone.
 - Using an additional immunosuppressant with prednisone may lower the dose of prednisone you need, which can lower the chance of prednisone side effects.

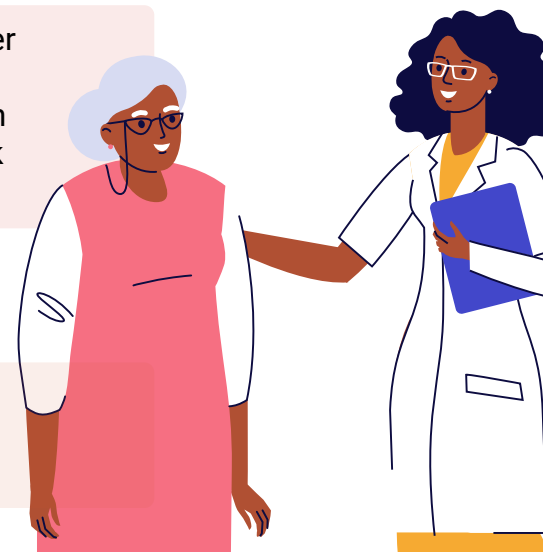
Treatment recommendations for refractory disease

If you have severe PAN that is refractory to prednisone and immunosuppressants other than cyclophosphamide:

- Switch the immunosuppressant drug to cyclophosphamide, over increasing prednisone alone.
 - We know that cyclophosphamide works well for people with newly diagnosed severe PAN, which suggests it would work well for refractory disease.

In general:

- We do not know the best length of time to take prednisone. How long you take it should be based on your condition and personal preferences.



Remission maintenance recommendations

If you have **newly diagnosed PAN that went into remission with cyclophosphamide:**

- ➔ Switch to another immunosuppressant, over continuing cyclophosphamide.
 - Cyclophosphamide should generally only be used for 3-6 months. Using it for longer can lead to serious side effects.



If you have **PAN in remission and are taking immunosuppressants:**

- ➔ Stop immunosuppressants after 18 months, over continued (indefinite) treatment.
 - Though people who have PAN sometimes relapse, many have only 1 episode of inflammation that does not return, so you may not need continued treatment.

Monitoring recommendations

If you have **severe PAN with belly area-related symptoms, but no longer have symptoms:**

- ➔ Get follow-up belly area vascular imaging to help track how well treatment is working.

If you have **PAN and peripheral neuropathy:**

- ➔ Get serial neurologic exams instead of repeated electromyography/nerve conduction studies to monitor the disease, because neurologic exams are less invasive (meaning they are procedures that do not enter your body).
 - When **would** I get electromyography/nerve conduction studies?
 - If you think you might have new neuropathy symptoms or your symptoms are getting worse.



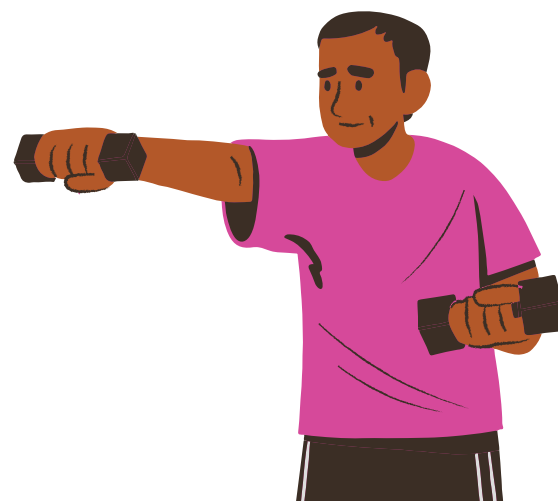
Other recommendations

If you have PAN with nerve and/or muscle-related symptoms:

→ We recommend physical therapy.

If you have PAN with symptoms of deficiency of adenosine deaminase 2 (DADA2):

→ Use tumor necrosis factor (TNF) inhibitors, over prednisone alone, because TNF inhibitors may be able to prevent strokes.



Health terms



- A**
 - **Abdominal vascular imaging:** Scans that create images of the blood vessels inside your belly area. Types of this imaging include:
 - **Conventional angiography:** A procedure used to see the blood vessels in your body. A catheter (a thin, flexible tube) is inserted into a blood vessel, and contrast material is injected through the catheter. Contrast material is a substance that makes your blood vessels more visible on an x-ray. An x-ray machine then takes a movie of how the contrast material moves through your vessels.
 - **Computed tomography (CT) angiography:** Uses multiple X-rays to make pictures of blood vessels.
 - **Magnetic resonance (MR) angiography:** Uses strong magnets to make pictures of blood vessels.
 - **Azathioprine:** An immunosuppressant. Lowers inflammation (swelling) in your body.
- B**
 - **Biopsy:** A procedure that removes a piece of your tissue so it can be examined under a microscope.
 - **Punch skin biopsy:** A round-tipped cutting tool is used to remove multiple layers of skin. A superficial punch biopsy removes fewer layers of skin than a deep or double biopsy, which removes more.
 - **Nerve biopsy:** Removal of a small piece of nerve.
 - **Muscle biopsy:** Removal of a small piece of muscle.
- C**
 - **Cyclophosphamide:** An immunosuppressant that lowers inflammation (swelling) in your body.
- D**
 - **Deficiency of adenosine deaminase 2 (DADA2):** A rare genetic disorder that causes inflammation (swelling), strokes, and skin changes.
- E**
 - **Electromyography/nerve conduction studies:** Tests that check how well muscles and nerves are working.
- I**
 - **Immunosuppressant:** A medicine that lowers your body's immune response to stop your immune system from causing inflammation (swelling) and damaging your body.
- M**
 - **Methotrexate:** An immunosuppressant. Lowers inflammation (swelling) in your body.
- P**
 - **Peripheral neuropathy:** Weakness, numbness, and pain caused by nerve damage, usually in your hands and feet.
 - **Plasma exchange:** A procedure that removes harmful materials in your blood by removing the plasma (the liquid part of blood) and replacing it with donor plasma or plasma substitute
 - **Prednisone:** A medicine that lowers inflammation (swelling) in the body, and can be given as:
 - **IV pulse:** A tube into a vein as an IV.
 - **Oral:** A pill by mouth.

S

- **Serial neurologic exams:** Exams that look for signs of neurologic damage which are repeated at each visit to track your progress over time. These exams may include testing your reflexes, looking for facial asymmetry, asking you to touch your nose with your eyes closed, and observing how you walk.

T

- **Tumor necrosis factor (TNF) inhibitors:** A medicine that lowers inflammation (swelling) in your body.

*Chung SA, Gorelik M, Langford CA, Maz M, Abril A, Guyatt G, Archer AM, Conn DL, Full KA, Grayson PC, Ibarra MF, Imundo LF, Kim S, Merkel PA, Rhee RL, Seo P, Stone JH, Sule S, Sundel RP, Vitobaldi OI, Warner A, Byram K, Dua AB, Husainat N, James KE, Kalot M, Lin YC, Springer JM, Turgunbaev M, Villa-Forte A, Turner AS, Mustafa RA. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Polyarteritis Nodosa. Arthritis Care Res (Hoboken). 2021 Aug;73(8):1061-1070. doi: [10.1002/acr.24633](https://doi.org/10.1002/acr.24633). Epub 2021 Jul 8. PMID: 34235889.

