POLYARTERITIS NODOSA (PAN)

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What is PAN?

PAN is a type of vasculitis affecting the medium-sized blood vessels in the body. Previously, PAN was commonly associated with hepatitis B infection, however with further understanding of these conditions, hepatitis B-associated vasculitis is now considered a separate entity from PAN.

Which organs are affected?

PAN most commonly affects the skin, kidney arteries causing infarcts, nervous system, joints and/or gastrointestinal tract. Heart involvement is possible but less common. The lungs are not affected.

Who is affected?

PAN is a rare condition affecting less than 30 per million people. Men are affected more than women. The typical age of disease onset is between 40 to 60 years.

Are there different forms of PAN?

PAN can be **systemic** (affects multiple organs) or **limited** to a specific organ.

PAN that is limited to the skin is called "cutaneous PAN". 10% of cutaneous PAN evolve into the systemic form. Another form is the genetically determined PAN, which includes **DADA2** (deficiency of adenosine deaminase 2). DADA2 is seen in young people and can cause strokes and a "PAN-like syndrome".

What are the clinical symptoms?

Depending on which organs are affected, PAN can cause:

- A painful lower extremity skin rash that can resemble "punched-out" wounds, or raised red nodules
- Prominent lacy discolouration of the blood vessels in the skin
- Nausea, abdominal pain or bloating, especially after eating
- Diffuse joint and muscle discomfort
- New or worsening hypertension
- Aneurysms and dilations of the arteries in the kidneys, liver or gastrointestinal tract
- Weakness, burning pain or tingling of the arms or legs
- Fevers, weight loss & unusually severe fatigue

How is PAN diagnosed?

Diagnosis is made based on a combination of the following:

1. Compatible symptoms and clinical features
2. Abnormal labs, including:
   - High inflammatory markers (ESR and/or CRP)
   - High creatinine and reduced kidney function
3. Supportive features on biopsy of affected organ(s). The most common site of biopsy is the skin.
4. Imaging of the abdominal area with vessel imaging (angiography) to assess for artery abnormalities
5. Cardiac imaging if there is suspected heart involvement
6. Electromyography if there is suspected nerve involvement

Diagnosing PAN may be challenging when the characteristic skin rash is absent.

Part of the diagnostic process is to rule out other diseases that disrupt the arteries. These may include other types of vasculitis, infections, malignancies, and rare genetic conditions.

N.B.: This information is intended for patient education, and for discussion with their physician(s). It is NOT a substitute for medical advice. Changes in treatment, based on this material, should always be reviewed with, and approved by, your physician(s). We encourage vasculitis patients to journal their progress, track their symptoms and know their medications, and lab and test results. © Vasculitis Foundation Canada June 2021 – **Last updated May 2021**

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Treatment of PAN includes:

- **Prednisone** – a steroid medication that is highly effective at reducing inflammation. You will start on a high dose and slowly reduce the dose over time.
- Other **immune suppressing medications,** which may be added immediately or later, once the prednisone dose is lower. These include methotrexate, azathioprine, leflunomide and mycophenolate mofetil. For severe or resistant disease, cyclophosphamide may be considered. For DADA2, anti-TNF agents are used.
- **Bone** and **stomach protectant** medications to prevent certain complications from high dose prednisone.
- **Bowel surgery** if the disease has led to intestinal tissue death or perforation.
- **Wound dressing** for deep skin ulcers.

The choice of immunosuppressive medication depends on the disease severity, which organs are affected, and whether you have other medical issues that prevent the use of certain medications.

How is my PAN monitored?

Your rheumatologist often works with other specialists to treat your PAN. Depending on which organs are affected by the disease, you may expect to see a **dermatologist**, **gastroenterologist**, or **neurologist**.

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 urine studies for blood and protein
- CT scans or MRI with angiography
- Echocardiogram (for the heart)
- Routine blood pressure monitoring

Testing is typically done more frequently at the beginning or when there is active disease, and less frequently once the disease is stable and in remission. Similarly, your visits with your specialists may be more frequent initially then become less frequent over time.

Can you survive PAN?

The survival rate for PAN is high when treated – up to 80% at 5 years. Survival depends on a variety of factors, including severity of disease, which organs are affected, and complications related to the disease or treatment. The most severe complications include heart attacks, strokes, bowel perforation, kidney failure and infections.

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

PAN is treatable but not yet curable. Currently, we do not know the optimal duration of treatment.

For most people, PAN follows a "monophasic" pattern, where there is a single episode of disease activity at the beginning without any disease recurrence or significant flares. These individuals may be able to eventually stop treatment.

Others, especially those with cutaneous PAN may experience more frequent flares (relapses) and need longer therapy.

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PAN DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Painful, red, bloodshot eyes
- ☐ Deep "punched-out" skin ulcers
- ☐ Raised painful skin nodules
- ☐ Burning, tingling or numbness of the hands or feet
- ☐ Weakness of the arm or leg
- ☐ Stroke symptoms
- ☐ Joint pain and/or joint swelling
- ☐ Muscle pain
- ☐ Abdominal or flank pain
- ☐ Bowel perforation
- ☐ Nausea and/or vomiting
- ☐ Bloody stools
- ☐ Fevers or drenching night sweats
- ☐ Weight loss
- ☐ Severe fatigue

Medications that have been prescribed:

	Date started	Date stopped
IV methylprednisolone _____ mg		
Prednisone: (Starting dose _____ mg)		
Cyclophosphamide _____ mg		
Septra/Bactrim/Sulfatrim		
Colchicine _____ mg		
Hydroxychloroquine _____ mg		
Methotrexate ____ mg		
Folic acid (only if taking methotrexate)		
Mycophenolate mofetil _____ mg		
Azathioprine ____ mg		
Leflunomide ____ mg		
Others:		

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

- ☐ Electromyography (EMG)
- ☐ X-rays (circle all that apply): Abdomen | Joints
- ☐ CT scans (circle all that apply): Abdomen | Head
- ☐ MRI (circle all that apply): Brain | Spine | Abdomen
- ☐ Ultrasound (circle all that apply): Abdomen | Joints
- ☐ Biopsy (circle all that apply): Skin | Bowel

What do I need to do?

- ✓ Attend **regular** follow-up visits with my rheumatologist and other specialists.
- ✓ Take my **medications** regularly and let my doctor know if I'm having side effects.
- ✓ **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.

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