PRIMARY ANGIITIS OF THE CENTRAL NERVOUS SYSTEM (PACNS)

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What is PACNS?

PACNS, or CNS vasculitis, is a rare immune mediated disease characterized by inflammation of the small- and/or medium-sized blood vessels in the brain. The vessels become narrowed and occluded, leading to brain ischemia (brain tissue injury due to inadequate blood supply), similar to what is seen in strokes.

Who is affected?

PACNS is extremely rare and affects men twice as much as women. Approximately 2-3 per million people are newly diagnosed every year, however we do not have enough data to know how many people in total have this disease. Most patients with PACNS are diagnosed in their 40-50s, but it can occur at almost every age.

What causes PACNS?

Like other autoimmune diseases, the specific cause of PACNS is still unknown. Certain infections and amyloid deposition (an abnormal form of protein) have been proposed as a trigger for the disease, however the specific details behind this are uncertain.

What are the symptoms?

- **Headache** – a slowly progressive and constant headache over several months is the most common symptom. A sudden onset severe ("thunderclap") headache should point your physician toward other diseases.
- **Cognitive change** – this includes noticeable memory loss, inability to perform usual daily tasks, and personality or behavioural changes. These changes are progressive over months, and beyond what is expected for healthy aging.
- **Recurrent strokes** – this includes sudden onset slurred speech, facial droop, inability to swallow, double vision or vision loss, dizziness, limb weakness, and in severe cases even paralysis.
- **Seizures**
- Loss of appetite & nausea
- Unusually severe fatigue

Symptoms are limited to the central nervous system and do not affect other organs such as the heart, lungs, skin, kidneys or joints. Involvement of other organs should point your physician toward other diseases.

How is PACNS diagnosed?

Diagnosis is made based on a combination of the following:

1. Compatible symptoms
2. Signs of inflammation in the spinal fluid – tested via a lumbar puncture
3. Imaging of the brain vessels with CT scan or MRI (or by conventional angiography)
4. Brain biopsy – this is the gold standard test (if the biopsy is positive then the diagnosis is definitive). However, results are often "falsely" negative, meaning that a negative biopsy does not definitively rule out the disease.

There are many diseases that can mimic PACNS, including other neurological or autoimmune diseases, other types of vasculitis, infections, cancers, atherosclerosis (due to cholesterol/plaque buildup) and drugs.

It is important to rule out these other conditions before making the final diagnosis. For this reason, PACNS can often take months to years to diagnose.

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Treatment of PACNS includes:

- **Prednisone** – this steroid medication is highly effective at reducing inflammation. You will start on a high dose and slowly reduce the dose over time.
- **Cyclophosphamide** – this immunosuppressant medication is often used during the first 3 to 6 months of treatment alongside prednisone. It can be given intravenously or by oral tablets.
- Other **immunosuppressant medications**, which are used after completing cyclophosphamide. These include methotrexate, azathioprine and mycophenolate mofetil. They can sometimes be used directly (without previous treatment with cyclophosphamide).

Adjunctive therapies include:

- **Blood-thinners** such as aspirin and/ or clopidogrel if you had strokes.
- **Bone and stomach protectant** medications to prevent certain complications associated with long-term prednisone use.
- **Pneumocystis pneumonia (PCP) protection** with an antibiotic if receiving certain immunosuppressive agents.

How is my PACNS monitored?

Your rheumatologist will typically work with a **neurologist** to treat your disease. A **neurosurgeon** may be involved early on to help perform the brain biopsy.

Your medical team (including your family physician) should also be monitoring you 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
- Repeat CT scans or MRI of the brain to assess for stability or progression of disease

Is a brain biopsy mandatory?

A biopsy needs to be considered for everyone with suspected PACNS to confirm the diagnosis. With that said, a brain biopsy is not without risks, so you or your doctors can be hesitant to go through with it. Empirical treatment would still be offered if the suspicion for PACNS is high enough, but with the understanding that the disease was not 100% confirmed.

What will happen to me?

Up to 80% of patients improve with treatment of the disease. The degree of functional and cognitive improvement depends on the severity of brain involvement and whether irreversible brain damage has occurred. Unfortunately, data on long-term disability is not yet available due to how rare the condition is. Around 1 in 4 patients will have a relapse of the disease.

How long do I need to stay on medications?

Like other autoimmune diseases, PACNS is treatable but not yet curable. Around 25% of individuals with PACNS will have a relapse of their disease. Currently, we do not know the optimal duration of treatment for PACNS.

Most individuals may expect to stay on immunosuppressive medication for at least 2 years. However, if you initially had severe disease and functional deficits, it may be more worthwhile for you to stay on long-term or even lifelong treatment to prevent a relapse. Stroke rehabilitation can also help with long-term functional recovery.

PACNS DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Headache
- ☐ Changes in memory
- ☐ Changes in personality
- ☐ Changes in behaviour
- ☐ Difficulty performing usual tasks
- ☐ Vision changes
- ☐ Weakness of the facial muscles
- ☐ Difficulty swallowing
- ☐ Arm weakness
- ☐ Leg weakness
- ☐ Seizures
- ☐ Vertigo (room spinning)
- ☐ Gait unsteadiness and recurrent falls
- ☐ Severe fatigue
- ☐ Others: _____

Medications that have been prescribed:

	Date started	Date stopped
IV methylprednisolone _____ mg		
Prednisone: (Starting dose _____ mg)		
Aspirin 81mg		
Clopidogrel 75mg		
Methotrexate ____ mg		
Folic acid (only if on methotrexate)		
Mycophenolate mofetil ____ mg		
Azathioprine ____ mg		
Leflunomide ____ mg		
Cyclophosphamide _____ mg		
Septra/Bactrim/Sulfatrim		
Others:		

I have had the following tests done:

- ☐ Blood tests checking for inflammation markers (ESR, and/or CRP)
- ☐ Lumbar puncture (spinal tap)
- ☐ CT scan of the brain
- ☐ MRI scan of the brain
- ☐ Conventional angiography
- ☐ Brain biopsy

What do I need to do?

- ✓ Attend **regular** follow-up visits with my rheumatologist and other specialists
- ✓ **Do regular** blood tests for disease and medication monitoring
- ✓ Ensure my medical team is monitoring me for diabetes, and control my blood pressure and cholesterol levels
- ✓ **Call 911 ASAP if I develop new stroke-like symptoms!**