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MADISON NEUROLOGY LLC

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Neurology Care 7 days a Week. Because Your Brain Matters.

CNS Vasculitis (Primary Angiitis of the Central Nervous System)

Patient education guide · Demyelinating & Autoimmune · Madison Neurology LLC

What is it?

CNS vasculitis is inflammation of the blood vessel walls within the brain and spinal cord, which narrows vessels and can cause strokes, bleeding, and widespread brain dysfunction. 'Primary' CNS vasculitis is confined to the nervous system; similar vascular inflammation can occur as part of systemic autoimmune diseases (lupus, rheumatoid arthritis, others). It is rare, serious, and treatable.

How does it happen?

The cause is unknown — the immune system attacks vessel walls, with inflammatory cells infiltrating and damaging them. Secondary forms accompany systemic autoimmune diseases, infections (varicella-zoster, others), or drug exposures. A look-alike condition, reversible cerebral vasoconstriction syndrome (RCVS), mimics it but behaves and is treated very differently.

Common symptoms

- Headache — often persistent and progressive
- Strokes or TIAs, sometimes multiple and in different brain regions
- Cognitive changes, confusion, personality change
- Seizures
- Vision problems or brain bleeding in some cases

How is it diagnosed?

Diagnosis is challenging and layered: MRI shows strokes or inflammation in multiple territories; vessel imaging (CTA/MRA/angiography) may show beading of arteries; spinal fluid shows inflammation. Brain biopsy is the definitive test and is performed when the diagnosis would change treatment. Extensive testing excludes infections and mimics such as RCVS, atherosclerosis, and lymphoma.

How is it treated?

Treatment is immunosuppression: high-dose corticosteroids, typically combined with cyclophosphamide or rituximab for induction, then maintenance agents (azathioprine, mycophenolate). Treatment usually lasts 1-2 years or more, with monitoring for both disease activity and medication effects. Stroke prevention measures and rehabilitation accompany immune therapy.

What does the future hold?

With treatment, most patients stabilize and many improve; untreated, the disease can be devastating. Relapses occur in a minority and respond to retreatment. Because diagnosis is difficult and treatment is potent, care at experienced centers matters.

Questions patients commonly ask

Q: How is this different from a regular stroke?

A: The strokes are caused by inflamed vessel walls rather than clots or cholesterol — so treatment targets the immune system, not just blood thinners.

Q: Why might I need a brain biopsy?

A: Because the treatment is long and strong, doctors want certainty; biopsy is the only test that directly proves vessel-wall inflammation.

Q: Is it related to lupus?

A: Primary CNS vasculitis is its own disease; lupus and other systemic conditions can cause similar brain vessel inflammation, which is why the full-body evaluation matters.

National support groups & resources

- **Vasculitis Foundation** — vasculitisfoundation.org
- **National Organization for Rare Disorders** — rarediseases.org — 1-800-999-6673
- **American Academy of Neurology — Brain & Life patient resources** — brainandlife.org

(Organization contact details were current at the time of compilation; please verify online.)

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