

Isolated central nervous system vasculitis also known as primary angiitis of the central nervous system (CNS), is a condition where the body's immune system attacks arteries of the brain, causing inflammation (swelling).

The **cause** of CNS is unknown. Researchers believe that some sort of event, like an infection, may trigger an inappropriate immune response directed mistakenly to the small and medium sized arteries of the brain.

Among the reported cases of CNS, there are more males who are diagnosed with the disease, and although most get the disease in their 40s or 50s, children as young as 3 and adults as old as 78 have been diagnosed with CNS.

The most common **symptoms** of CNS are: confusion, headache, personality changes, and muscle weakness/paralysis similar to what happens when someone suffers a stroke. Other symptoms include seizures, bleeding in the head, coma, and vision loss. Symptoms usually occur over the course of several months, but can also occur very quickly.

Nonspecific **laboratory abnormalities** have been noted among patients with CNS. The diagnosis of CNS can be done in different ways. The principal form of diagnosis remains angiography (x-rays of arteries), but magnetic resonance imaging (MRI) and CT scanning have also been utilized. None of these alone can be diagnostic of CNS. A common concern is that angiography in a patient with possible cerebral vessel inflammation is unsafe, and may result in complications. However a study of 125 cerebral angiograms, found no greater risk of complications in patients with proven CNS vasculitis than in those with normal angiograms. A lumbar puncture may also be performed to look for abnormalities in the cerebrospinal fluid.

The symptoms, signs, or findings among patients with CNS may "mimic" a number of disorders including other primary CNS vasculitides, systemic vasculitis, CNS infections or drug-induced disorders by methamphetamine, cocaine, "crack", and ephedrine abuse.

Early reports of patients with cerebral vasculitis suggested a less optimistic prognosis but more recent experience with immunosuppressive therapy has resulted in encouraging outcomes.

If the **diagnosis** of CNS is confirmed by biopsy, treatment with high doses of glucocorticoids (steroids) is warranted. Cytotoxic agents such as cyclophosphamide (Cytoxan) can be added depending on the clinical severity of the vasculitis.

Side-effects of high-dose glucocorticoids should be anticipated such as glucose intolerance or frank diabetes mellitus (condition of abnormal glucose levels in blood). Use of calcium supplements, vitamin D, hormone replacement therapy, and bisphosphonates can be taken to help protect the body from bone mineral loss. Antibiotics can also be taken to help prevent infection due to the use of immunosuppressants.