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CADASIL (Cerebral Autosomal Dominant Arteriopathy with Sub-cortical Infarcts and Leukoencephalopathy)

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). <u>CADASIL Information</u> <u>Page.</u>

CADASIL (Cerebral Autosomal Dominant Arteriopathy with Sub-cortical

Infarcts and Leukoencephalopathy) is an inherited form of cerebrovascular disease that occurs when the thickening of blood vessel walls blocks the flow of blood to the brain. The disease primarily affects small blood vessels in the white matter of the brain. A mutation in the Notch3 gene alters the muscular walls in these small arteries. CADASIL is characterized by migraine headaches and multiple strokes progressing to dementia. Other symptoms include cognitive deterioration, seizures, vision problems, and psychiatric problems such as severe depression and changes in behavior and personality. Individuals may also be at higher risk of heart attack. Symptoms and disease onset vary widely, with signs typically appearing in the mid-30s. Some individuals may not show signs of the disease until later in life. CADASIL - formerly known by several names, including hereditary multi-infarct dementia — is one cause of vascular cognitive impairment (dementia caused by lack of blood to several areas of the brain). It is an autosomal dominant inheritance disorder, meaning that one parent carries and passes on the defective gene. Most individuals with CADASIL have a family history of the disorder. However, because the genetic test for CADASIL was not available before 2000, many cases were misdiagnosed as multiple sclerosis, Alzheimer's disease, or other neurodegenerative diseases.