Cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)

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Proposed Diagnostic Criteria for Cerebral Autosomal-Dominant Arteriopathy With Subcortical Infarcts and Leukoencephalopathy

1. Probable cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL):

   a. Young age at onset (≤50 years of age).

   b. At least two of the following:

      i. Clinical stroke-like episodes with permanent neurological signs.

      ii. Migraine.
iii. Major mood disturbances.

iv. “Subcortical-type” dementia.

c. No vascular risk factor etiologically related to the deficit.

d. Evidence of an inherited autosomal-dominant transmission.

e. Abnormal magnetic resonance imaging (MRI) imaging of the white matter without cortical infarcts.

2. Definite CADASIL:

a. Criteria of probable CADASIL associated with linkage to NOTCH 3 mutation, and/or

b. Pathological findings demonstrating small vessel arteriopathy with granular osmiophilic material.

3. Possible CADASIL:

a. Late age at onset (≤50).

b. Stroke-like episodes without permanent signs, minor mood disturbances, global dementia.

c. Minor vascular risk factors, such as mild hypertension, mild hyperlipidemia, smoking, and/or use of oral contraceptives.

d. Unknown or incomplete family pedigree.

e. Atypical MRI imaging of the white matter.
4. Exclusion criteria:

a. Age at onset over 70 years.

b. Severe hypertension or complicated heart or systemic vascular disease.

c. Absence of any other case in a documented pedigree.

d. Normal MRI imaging, age over 35 years.

Reference:


Category: Cerebrovascular disorders Diagnostic Criteria Tags: CADASIL diagnostic criteria

See also:

1. NINDS-AIREN Criteria for the Diagnosis of Vascular Dementia
2. DSM-IV Criteria for the Diagnosis of Vascular Dementia
3. Probable Alzheimer’s Disease According to NINCDS-ADRDA Criteria