Cerebral Cavernous Malformation Classification

- Natural History of Sporadic Cerebral Cavernous Malformations by Zabramski Classification: Hemorrhage Risk and Functional Outcomes Over 5 Years
- Auto-segmentation of cerebral cavernous malformations using a convolutional neural network
- Clinical, genomic, and histopathologic diversity in cerebral cavernous malformations
- Clinical and radiological presentation of cavernomas according to the Zabramski classification
- Neurological and functional outcomes of 32 patients with hemorrhagic brainstem cavernous malformations: a practical guide for surgical planning
- A taxonomy for cerebellar cavernous malformations: subtypes of cerebellar lesions
- Cavernous Malformation From Cranial Nerves: A Systematic Review With a Novel Classification and Patient-Level Analysis
- Pediatric Central Nervous System Vascular Malformation : Pathological Review with Diagram

Cerebral cavernous malformations (CCMs) can be classified based on histopathology, genetics, radiological features, clinical presentation, and anatomical location.

1. Histopathological Classification

CCMs are vascular lesions composed of clusters of dilated, thin-walled capillaries without intervening brain parenchyma. They lack normal vessel structure, with no smooth muscle or elastic tissue.

Zabramski Classification (1994)

see Zabramski Classification

2. Genetic Classification

CCMs can be **sporadic** or **familial (autosomal dominant with incomplete penetrance)**.

- **Sporadic CCMs**: Usually solitary, often associated with developmental venous anomalies (DVAs).
- **Familial CCMs**: Multiple lesions with a higher risk of recurrence due to genetic mutations.

see Familial cerebral cavernous malformation.

Sporadic CCMs occur in people with no family history of the disorder. These individuals tend to have only one CCM. Those with sporadic CCM do not have a greater chance of having a child with a CCM than anyone else in the general population.

Three main genes are implicated:

- **CCM1 (KRIT1)** – Chromosome 7q21.2

- **CCM2 (MGC4607)** – Chromosome 7p15-13
- **CCM3 (PDCD10)** – Chromosome 3q26.1

3. Clinical Classification

- **Asymptomatic**: Incidental findings on imaging.
- **Symptomatic**:
  1. **Seizures**: Most common presentation.
  2. **Hemorrhagic**: Acute or chronic bleeding leading to focal neurological deficits.
  3. **Progressive neurological deterioration**: Due to recurrent hemorrhages or mass effect.

4. Location-based Classification

- **Supratentorial**: Commonly associated with seizures.
- **Infratentorial**: Can lead to brainstem dysfunction.

This classification system helps in assessing prognosis, determining treatment strategies, and understanding genetic risks.

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see Insular Cavernous Malformation

see Frontal lobe cavernous malformation.

see Temporal lobe cavernous malformation.