



Reversible Cerebral Vasoconstriction Syndromes

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Reversible cerebral vasoconstriction syndromes (RCVS) show reversible multifocal narrowing of the cerebral arteries with clinical manifestations that characteristically include thunderclap headache and less commonly focal neurologic deficits related to brain edema, stroke, or seizure. RCVS predominantly affects women (female to male ratio 2:1 to 10:1, depending on the case series). The mean age across published studies is 42 to 44 years. RCVS has been associated with a multiplicity of conditions including pregnancy, migraine, use of vasoconstrictive drugs and other medications, neurosurgical procedures, hypercalcemia, unruptured saccular aneurysms, cervical artery dissection, cerebral venous thrombosis, and others. Between 30 and 70 percent of patients with RCVS have no abnormality on initial brain scans, despite having widespread cerebral vasoconstriction. However, approximately 75 percent of admitted patients eventually develop parenchymal lesions. The most frequent lesions are ischemic stroke and cortical surface (nonaneurysmal or convexal) subarachnoid hemorrhage, followed by reversible vasogenic brain edema and parenchymal hemorrhage. Infarcts are often bilateral and symmetrical, located in arterial "watershed" regions of the cerebral hemispheres or in the cortical-subcortical junction. Larger infarcts are often wedge-shaped. Cerebral angiographic abnormalities are dynamic and progress proximally, resulting in a "sausage on a string" appearance of the circle of Willis arteries and their branches. There is no proven therapy for RCVS. Supportive care is directed towards managing blood pressure, severe headaches, and other complications such as seizures. Oral calcium channel blockers are often administered to treat vasoconstriction but the supporting evidence for this strategy is weak.

Key words: RCVS, stroke



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