Moyamoya syndrome

- Development and validation of a predictive model for perioperative low-density lipoprotein as a risk factor for postoperative cerebral infarction in Moyamoya disease
- The Differences of Cerebral Hyperperfusion in Patients With Moyamoya Disease and Atherosclerotic Occlusive Cerebrovascular Disease Evaluated by 99mTc-ECD SPECT One Day After Revascularization Surgery
- The Orbital Grading system yields higher precision than the Matsushima grading system in assessing angiographic outcomes after EDAS for Moyamoya disease: an interrater reliability analysis
- Moyamoya disease in early infancy: Report of youngest Indian infant
- Long-Term Outcomes in Patients With Hemorrhagic Moyamoya Disease Combined With Hypertension After Encephaloduroarteriosynangiosis
- Association of intravascular enhancement sign on 3D T1- weighted TSE sequences with cerebral perfusion and infarction events in moyamoya disease
- Blurred by a "Puff of Smoke"-A Case-Based Review on the Challenging Recognition of Coexisting CNS Demyelinating Disease and Moyamoya Angiopathy
- Recent Advances in Genetics of Moyamoya Disease: Insights into the Different Pathogenic Pathways

Feature	Moyamoya Disease (MMD)	Moyamoya Syndrome (MMS)
Cause	Idiopathic / primary	Secondary to another condition
Laterality	Typically bilateral	Can be unilateral or bilateral
Associated conditions	None	Yes (e.g., NF1, Down syndrome, sickle cell, lupus)
Management	Surgical revascularization	Surgical + treat underlying cause
Diagnostic label	Used when no other cause is found	Used when an associated condition is identified
Imaging appearance	“Puff of smoke” vessels on angiography	Same appearance as MMD

Moyamoya syndrome can be associated with other medical conditions such as sickle cell disease, neurofibromatosis, Down syndrome, autoimmune diseases, radiation therapy, and other vascular disorders. Acquired: Unlike Moyamoya disease, Moyamoya syndrome is often acquired as a result of another health issue or external factor.

Case series

Yang et al. evaluated the value of high-resolution magnetic resonance imaging of the vessel wall (VWI) for differentiating moyamoya disease (MMD) from atherosclerotic moyamoya syndrome (AS-MMS).

Materials and methods: Twenty-one patients with MMD or AS-MMS were assessed retrospectively by two independent raters regarding and magnetic resonance angiography (MRA) stage grading score; collateral development in the lateral fissure and basal ganglia on MRA; and pattern of the thickening

of the arterial wall; presence, degree, and pattern of enhancement; presence and distribution of deep tiny flow voids (DTFVs) and collateral development in the lateral fissure and basal ganglia on VWI. After univariate analysis between the two groups, logistic regression models based on imaging findings of MRA or VWI were implemented respectively, and receiver operating characteristic (ROC) curves were generated to compare the discriminatory power of the two imaging methods for diagnosis of MMD. Interrater agreement was analysed using an unweighted Cohen's κ or interclass correlation coefficient (ICC).

Results: MMD manifested as more concentric thickening, more homogeneous enhancement, higher presence of DTFV, smaller outer-wall boundary area of stenosis or occlusion, and smaller remodelling index on VWI. After Bonferroni-Holm correction for multiple comparisons, for AS-MMS, collaterals in both the lateral fissure and basal ganglia were not usually present on either MRA or VWI. The diagnostic performance of the multivariate logistic regression model based on VWI with an accuracy of 87.1% for classification was higher than MRA. Interrater agreement was moderate or substantial for all the imaging findings.

High-resolution magnetic resonance imaging of the vessel wall (VWI) might be a useful and feasible method for differentiating moyamoya disease (MMD) from atherosclerotic moyamoya syndrome (AS-MMS) and a prospective tool for guiding first-line treatment [1].

Case reports

A 7-year-old boy with Down syndrome and atlanto-axial subluxation. The patient presented with an ischemic stroke in the left hemisphere and cervical cord compression with increased cord edema. Diagnostic digital subtraction angiography revealed unique patterns of vascular involvement, with retrograde flow through the anterior spinal artery, ascending cervical artery, occipital artery, and multiple leptomeningeal arteries compensating for bilateral vertebral artery occlusion. This case underscores the underreported phenomenon of upward retrograde flow through the anterior spinal artery in bilateral vertebral artery occlusion. They address the rare manifestation of posterior circulation involvement in moyamoya syndrome, highlighting the importance of considering atlantoaxial instability as a contributing factor, as the absence of atlantoaxial stability is a risk factor for vertebral artery dissection. This study contributes valuable insights into the intricate relationship of moyamoya syndrome, Down syndrome, and atlantoaxial instability, urging clinicians to consider multifaceted approaches in diagnosis and treatment. It also emphasizes the potential significance of the anterior spinal artery as a compensatory pathway in complex vascular scenarios [2]

1)
Yang S, Wang X, Liao W, Li L, Tan Z, Zhu L, Hu P, Cui X, Xing W. High-resolution MRI of the vessel wall helps to distinguish moyamoya disease from atherosclerotic moyamoya syndrome. Clin Radiol. 2021 Feb 11:S0009-9260(21)00018-0. doi: 10.1016/j.crad.2020.12.023. Epub ahead of print. PMID: 33583567.
2)
Abramyan A, Fu AY, Patel K, Sun H, Roychowdhury S, Gupta G. Neurovascular considerations in patients with Down syndrome and moyamoya syndrome. Childs Nerv Syst. 2024 Jan 25. doi: 10.1007/s00381-024-06293-z. Epub ahead of print. PMID: 38273142.