Moyamoya: angiographic findings of unilateral/ bilateral stenosis or occlusion of the arteries around the circle of Willis **WITH** prominent arterial colateral circulation

Moyamoya Disease (MMD) - Primary/idiopathic

Primary/ Idiopathic Moyamoya Disease

Genetic associations

- RNF213 gene mutation in Eastern Asian populations
- Locus 17q25 mutation
- Novel Risk Foci
  - Homocysteine Metabolism Regulation Genes
  - Large Vessel Disease Associated Foci

Moyamoya Syndrome (MMS) - secondary form

Associated condition

Diseases affecting arteries around the Circle of Willis

- Atherosclerosis
- Cranial Radiation
- Cranial Trauma
- Brain Tumors
- Meningitis

Hematologic Conditions

- Sickle Cell Disease
- Beta- thalassemia
- Fanconi Anemia
- Homocystinuria/ hyperhomocysteinemia
- Factor XII deficiency
- Essential Thrombocytopenia
- Protein S deficiency
- Pyruvate Kinase Deficiency

Genetic Disorders

- Alagille Syndrome
- Down Syndrome
- Hypomelanosis of Ito
- Marfan Syndrome
- Microcephalic Osteodysplastic Primordial Dwarfism Type 2
- Neurofibromatosis Type I
- Noonan Syndrome
- Phakamatosis Pigmentovascularis type IIIb
- Prader-Willi Syndrome
- Pseudoxanthoma elasticum
- Sturge-Weber Syndrome
- Tuberous Sclerosis
- Turner Syndrome
- Williams Syndrome

Vasculitis/ Autoimmune/Multisystem diseases

- SLE
- Polyarteritis nodosa and postinfectious vasculopathy
- Graves Disease/ thyroiditis
- Sneddon Syndrome/ Antiphospholipid Antibody Syndrome
- Anti-Ro / Anti-La Antibodies
- Type 1 Diabetes Melitus
- Pulmonary Sarcoidosis

Other Vasculopathies

- Coarction of the Aorta
- Congenital Heart Disease
- Fibromuscular Dysplasia
- Renal Artery Stenosis

Metabolic Disease

- Type I Glycogenosis
- Hyperphosphatemia
- Primary Oxalosis

Renal Disorders

- Polycystic Kidney Disease
- Wilms tumor

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