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MOYAMOYA DISEASE

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ABSTRACT

Moyamoya disease is a disease in which arteries in the brain are constricted. Moyamoya disease is first identified in Japan. Moyamoya means "puff of smoke" in Japanese.

Keywords: Moyamoya Disease, internal carotid artery, Ischemia, Epilepsy, Hemiparesis, Magnetic Resonance Angiography, Revascularization

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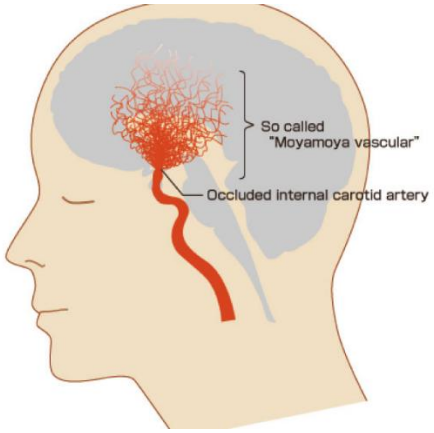
INTRODUCTION

It is a rare, chronic and progressive cerebrovascular disorder caused by narrowing and blocking of internal carotid artery. Carotid artery is the artery carry oxygenated blood to brain.

DEFINITION

Moyamoya disease is an isolated, chronic usually bilateral, vasculopathy characterized by progressive narrowing of the terminal intracranial portion of the internal carotid artery and Circle of Willis



INCIDENCE

- Often diagnosed in children 10-14 years old or in older adults in their 40's
- Female have highest incidence compare to male
- East Asian ethnicity having highest incidence

CAUSES

- Exact causes are unknown
- Studies says that greater prevalence in Asian countries strongly suggest a genetic factor, changes in one gene RNF213
- Family history – the risk of having condition is 30 to 40 times higher than other general population
- Associated with other conditions like
  - Down Syndrome
  - Sickle Cell Anaemia
  - Neurofibromatosis type I
  - Hyperthyroidism

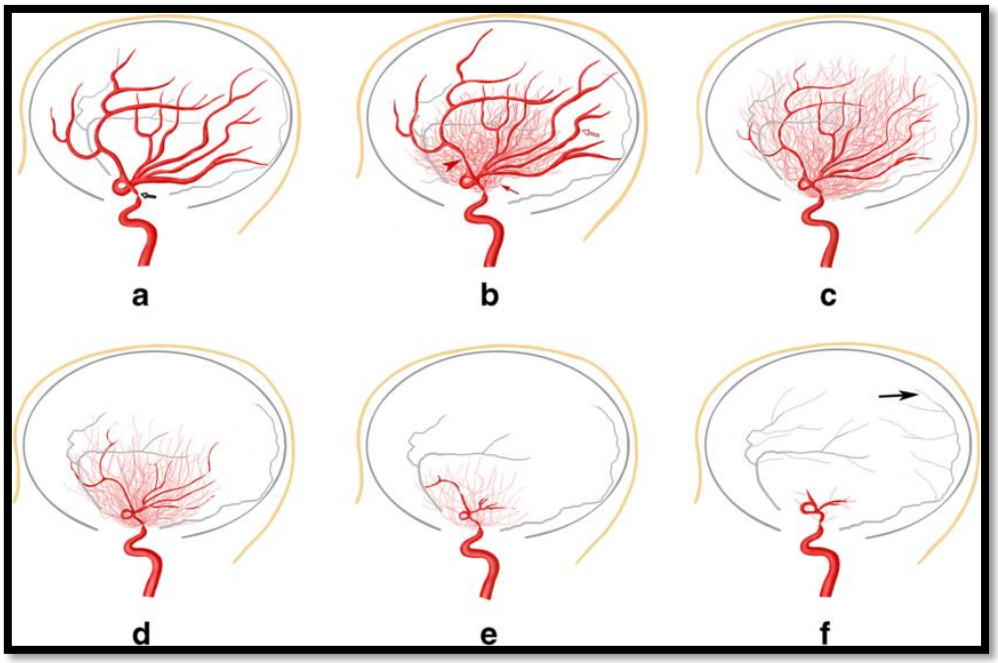
SYMPTOMS

- Recurrent Transient Ischemia Attack
- Epilepsy
- Stroke – Ischemic Stroke or haemorrhagic Stroke
- Hemiparesis
- Progressive difficulty in thinking and remembering
- Related to decreased blood flow
  - Headache
  - Seizures
  - Weakness, numbness or paralysis in face, arm or leg
  - Vision problem
  - Trouble speaking or understanding others
  - Cognitive and developmental delays
  - Involuntary movements

DIAGNOSIS

- History of the patient
  - Recurrent ischemic strokes
  - Family history
  - Associated conditions
- Physical examination
- MRI – the amount of blood passing through the vessels, reduction of blood supply to brain
- MRA (Magnetic Resonance Angiography) – degree of narrowing
- CCA (Conventional Cerebral Angiography) – provide accurate information about the area and degree of narrowing
- According to Suzuki system it can be classified into 6 stages

SATGES	**DESCRIPTION**
1	Narrowing of the terminal internal carotid artery
2	Initiation of moyamoya vessels in basal carotid circulation, dilation of intracerebral arteries
3	Intensification of moyamoya vessels, severe carotid stenosis, defection of anterior cerebral artery and middle cerebral artery
4	Minimization of the moyamoya and defects to the posterior cerebral artery
5	Reduction of the moyamoya and development of external carotid collateral
6	Disappearance of the moyamoya and circulation only through external carotid artery and vertebral artery



- Transcranial Doppler (TCD) – provide blood flow velocity
- Electro Encephalography (EEG)
- Positron Emission Tomography (PET) scan or Single Positron Emission Computerized Tomography (SPECT) – blood flow to regions of the brain

TREATMENT

- Goal:
  - Reduce symptoms and improve brain blood flow
  - Lower the risk of complications
- Conservative Management
  - Antiplatelet drugs – e.g Aspirin
  - Analgesics
  - Antiepileptic drugs
- Surgical Management
  - Direct Revascularization procedure: superficial temporal artery to middle artery bypass surgery
  - Indirect Revascularization: easier method but the time to improve the cerebral blood flow is longer than direct vascularization. Major technique used under these methods are
    1. Encephalomyo Synangiosis (EMS)
    2. Encephalo-duro-arterio synangiosis (EDAS)

PROGNOSIS

Overall prognosis is variable. Two-thirds of patient with disease have a symptomatic progression over 5 years with poor outcomes. Early surgical revascularization has good prognosis.

COMPLICATIONS

- Intraoperative ischemic stroke
- Post operative ischemic stroke with permanent neurologic defect (0.9 to 8% of patients)
- Haemorrhagic shock (0.7 to 8% of patients)
- Postoperative epidural hematoma (4-8% of paediatric patients)
- Hyper perfusion syndrome after direct vascularization (21.5 to 50% of patients)
- Scalp ischemia (17.6 to 21.4% of patients)

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