Moyamoya Disease:

- Chronic cerebrovascular disease of **unknown aetiology** characterised by **bilateral** progressive stenosis and occlusion of the distal ICA and proximal ACA and MCA and the development of **secondary** collateral pathways to compensate for the decrease in regional cerebral blood flow

- Rare disease described mainly in Japanese and Asian population. Some cases reported in other ethnic groups. Incidence in Japan 1: 1000000 per year. M:F 1: 1.6
- Two modes of presentation:

1. **In children** (95%-**ischemic episodes** either transient ischemic attacks or **permanent stroke**, motor sensory or visual involving different parts of the body bilaterally precipitated by **hyperventilation**. 5% with haemorrhage).
2. In **Adults**-Haemorrhage (SAH, ICH) from dilated collaterals, pseudoaneurysms on the basal collaterals and aneurysms which are present in 1% of children and 6% of adults.

- Pathology: **primary event** is progressive stenosis of supraclinoid ICA due to intimal hyperplasia and folding of internal elastin lamina (b-FGF in the intima?, decrease receptors for PDGF in myocytes) with **secondary** development of collaterals

1. **Basal perforating collaterals**(branches of posterior cerebral artery (thalamoperforators) and ACA and MCA (medial and lateral lenticulostriate artery), tentorial branches of ICA, anterior ethmoidal and posterior ethmoidal branches of OA, mamillary, interpeduncular, medial and lateral posterior choroidal branches of PCA)
2. **Leptomeningeal collaterals** (between posterior and anterior circulation)
3. **Dural vessels** can develop anastomoses with pial vessels (MMA, accessory meningeal artery)
4. **External carotid branches** (occipital artery, STA, Maxillary artery - eventually develop transdural anastomosis). This is the most rich anastomosis and usually coincides with spontaneous resolution of symptoms

- Diagnosis:

1. CT scan : areas of infracts, gyral enhancement post contrast, Ivy sign (dilated collaterals)
2. MRI, MRA: 75% sensitivity, 100% specificity in diagnosing the condition
3. Cerebral angiogram including ECA. (puff of smoke)
4. SPECT, PET, Xenon CT scan- regional cerebral blood flow
5. EEG- Centro temporal slowing with re build phenomenon(slow waves with high amplitude produced by hyperventilation and lasting 10 min =cortical ischemia

- Diagnostic criteria:

1. Bilateral stenosis or occlusion of supraclinoid ICA
2. Basal and Leptomeningeal collaterals on angiogram (puff of smoke)
3. No identifiable causes(atherosclerosis, SLE, NF and so on)

If the disease is unilateral and there is underlying cause, it is called Moyamoya syndrome or variant

1. Treatment: surgery is indicated for patients with recurrent ischemic symptoms and for clipping of associated aneurysms. **During anaesthesia avoid hypotension, hypovolemia and hypocapnia.**

1. **Direct revascularisation procedures**(STA-MCA bypass) is out of favour(disruption of naturally occurring collaterals, technical difficulty and the need to occlude the MCA branch during the anastomosis
2. **Indirect revascularisation**

A. EMS (encephalomyosynagiosis). Placing vascular temporalis muscle on the pial surface. Risk of bleeding, focal seizures, large bony defect

B. EDAS (encephalodurosynangiosis). Resolution of symptoms in **75% at 1 year and 97% at 2 years**

C. Pial synangiosis (suturing STA to the arachnoid).

2. Anaesthetic precautions(avoid hypocapnia, hypotension and hypovolemia)