

# A BRIEF REVIEW ON MOYAMOYA DISEASE

G. Naga Chandrika, Dr. V. Asha Jyothi

Department of Pharmacy, St. Pauls college of pharmacy, Turkyamjal, Hyderabad, Telangana.

## ABSTRACT:

Moyamoya is a rare cerebrovascular disease. It affects both adults and paediatrics. It frequently occurs in east Asian populations. Symptoms include ischemic or haemorrhagic stroke, head ache, epilepsy. It is diagnosed mainly by using cerebral angiography. Pathophysiology is not clear but due to some genetic mutations this disease occurs. Normally it is treated by surgery.

**KEYWORDS:** Angiogenesis, Caveolin, Growth factors, Moyamoya disease.

## INTRODUCTION:

Moyamoya disease is a rare cerebrovascular disease. It is characterized by steno occlusive change at the end of the internal carotid artery, middle cerebral artery, which is accompanied by the formation smoke like abnormal blood vessels in the base of the skull in digital subtraction angiography. <sup>(1)</sup> By using cerebral angiography reveals smog like blood vessels in the base of the skull<sup>(2)</sup>.

## EPIDEMIOLOGY:

The incidence of Moyamoya disease is more in east Asian countries but low in European countries. In foreign countries females more are affected than the males. <sup>(3)</sup> Etologically the exact reason for the disease is unknown.

## PATHOPHYSIOLOGY:

The basic pathology of Moyamoya disease mainly includes intimal fibrous hyperplasia of the intracranial arterial stenosis, irregular proliferation of the inner elastic layer, thinning of the middle layer of the vessel wall and reduction of the outer diameter of the blood vessel. <sup>(4)</sup>

Deficiency or low frequency variation Of Ringin protein 213(RNF 213) P.R4828K causes mutation of this protein on exon 61 and exon 60 causes abnormality in the development of abnormal vascular network. <sup>(5)</sup>

Caveolin 1is plasma membrane protein. Decreased serum levels of caveolin1 leads to decrease in the RNF213.

<sup>(6)</sup>

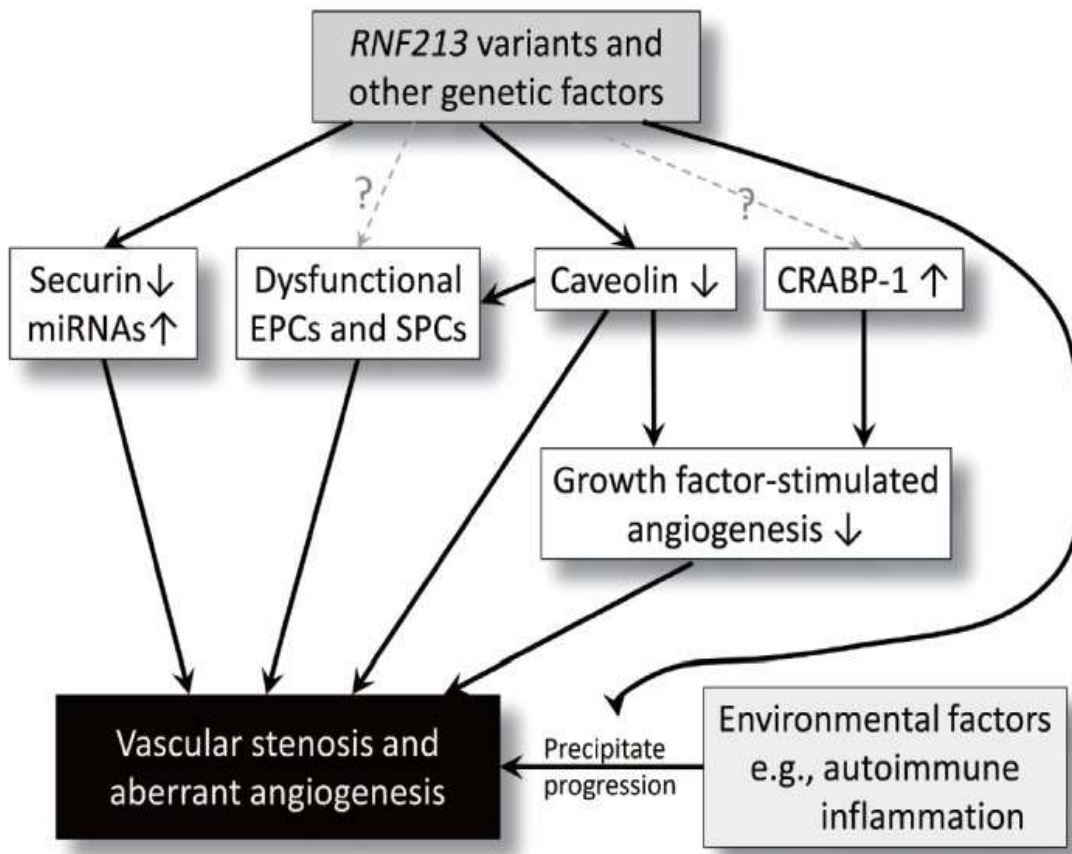


Fig. 1: Pathophysiology of Moya Moya disease.<sup>8</sup>

### SYMPTOMS:

- Ischemic stroke
- Hemorrhagic stroke
- Head ache
- Cerebral ischemic
- Epilepsy
- Cognitive dysfunction

### DIAGNOSIS:

- Cerebral angiography:
  - stages and cerebral angiographic findings.
  - Stage I-Arrowing of the carotid fork.
  - Stage II-Initiation of the Moyamoya.
  - Stage III-Intensification of the Moyamoya.
  - Stage IV-minimization of the Moyamoya.

Stage V-Reduction of the Moyamoya.

Stage VI-Disappearance of the Moyamoya.

- Poison emission tomography (PET)
- Single photo emission computed tomography <sup>(7)</sup>.
- MRA

#### TREATMENT:

There is no specific treatment for this disease. Mainly the symptoms of this disease by giving Antiplatelets, Anti coagulants. Mainly it is treated by surgery only. There are 3 types:

- Direct revascularization
- Indirect revascularization
- Combined revascularization <sup>(7)</sup>.

#### CONCLUSION:

Moyamoya is the rarest disease, such that more research must be conducted on etiology, pathophysiology. At present the treatment is given based on symptoms presented, extended the treatment based on the diagnosis.

#### REFERENCES :

1. Takeuchi. K, Shimizu. K, Hypoplasia of the bilateral internal carotid arteries. Brain Nerve. 1957; 9: 37 - 43.
2. Suzuki. J, Takaku. A, Cerebrovascular Moyamoya disease. Disease showing abnormal net like vessels in base of brain. Arch Neurol. 1969; 20; 288-299.
3. Zhou. X. W, Wang. J. Z, Ma. Y. Literature review and current situation of domestic Moyamoya disease. Chin J Pract Nerv Dis. 2017; 20:1-4.
4. Kuroda. S, Houkin. K. Moyamoya disease: current concepts and future perspectives. Lancet Neurol. 2008; 7: 1056 - 1066. doi. 10. 1016 / S1474 - 422 (08) 70240.
5. WuZ, Jiang. H, Zhang. L, X. U. X, Zhang. X, Kang. Z, Song. D, Zhang. J, Guan. M, Gu. Y. Molecular analysis of RNF 213 gane for Moyamoya disease in the Chinese Han population. Plos one 2017;7: e48179. doi: 10.1371 / journal. pone. 0048179.
6. Liu. J, Wang. X.B, Park. D.S, Lisanti. M.P. Caveolin-1 expression enhances endothelial capillary tubule formation. The journal biological chemistry. 2002; 277: 10661 - 10668.
7. Lee. S. U, Oh. C.W, Kwon. O. K, Bangs. Js, Ban. S. P, Byoun. H. S, Kim. T. Surgical treatment of adult Moyamoya disease. Curr treat options Neurol.2018; 20: 22. doi: 10. 1007 / s11940 – 018 – 0511 - 8.
8. Journal of Stroke 2016;18(1):12-20 <http://dx.doi.org/10.5853/jos.2015.01760>.