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Moyamoya Disease: Prognosis, Evaluation and Therapy

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Short Communication

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DESCRIPTION

A condition known as Moyamoya disease causes narrowing of some brain vessels. Blood flow is blocked by constriction and blood clots. Around the blocked vessels, a collateral circulation develops to compensate for the blockage, but these collateral vessels are weak, small, and prone to bleeding, aneurysms, and thrombosis. These collateral vessels appear as a "puff of smoke" on conventional angiography, which is called "Moyamoya" in Japanese. Moyamoya is identified as moyamoya illness when it occurs all by itself and has no underlying correlating diseases. This also holds true when bilateral collateral circulation and arterial constriction are present. Unilateral arterial constriction, or moyamoya syndrome, happens when one of the several disorders listed also exists. This might also be thought of as the primary condition's secondary manifestation, or moyamoya. The distal internal carotid artery is primarily blocked. When angiography reveals a "puff of smoke" appearance, surgical bypass is the preferred course of action [1].

Moyamoya disease affects about 10% of people in families, and some cases are caused by particular genetic abnormalities. Variation in the RNF213 gene (613768) on the long arm of chromosome 17 confers susceptibility to moyamoya disease-2 (MYMY2; 607151). The ACTA2 gene (102620) on the long arm of chromosome 10 (10q23.3) is the source of moyamoya disease-5 (MYMY5; 614042), while the GUCY1A3 gene (139396) on the long arm of chromosome 4 is the source of moyamoya disease-6 with achalasia (MYMY6; 615750). The short arm of chromosome 3 (MYMY1) and the long arm of chromosome 8 (8q23) have both been identified as loci for the condition (MYMY3; 608796). X-linked recessive syndromic condition MYMY4 (300845), which is characterized by moyamoya disease, small stature, short stature, hyper gonadotropic hypogonadism, and facial dysmorphism [2].

The incidence rate of moyamoya in the US is 0.086 per 100,000. Overall incidence is greater in Japan (0.35 per 100,000). Compared to patients from Japan, they are more likely to experience repeated strokes and may be dealing

with a unique underlying pathophysiology. Both congenital and acquired moyamoya disease are possible. Moyamoya malformations can occur in people with down syndrome, Sickle cell anemia, Neurofibromatosis type 1, Congenital heart disease, Fibromuscular dysplasia, Activated protein C resistance, or Head trauma [3].

The gold standard for determining the progression of moyamoya disease is cerebral angiography. It can be divided into six stages using Suzuki's classification system.

Stage 1: Carotid fork narrowing

Stage 2: Moyamoya initiation and enlargement of the cerebral major arteries

Stage 3: Moyamoya intensification and abnormalities in the middle and anterior cerebral arteries

Stage 4: Moyamoya minimization and posterior cerebral artery problems

Stage 5: Moyamoya reduction and formation of collaterals in the external carotid artery

Stage 6 Moyamoya disappears and circulation is limited to the vertebral artery and external cerebral artery.

Studies in nuclear medicine, such as Single Photon Emission Computerized Tomography (SPECT), are frequently used to show how the brain's affected regions receive less blood and oxygen. Prior to considering any surgical options, conventional angiography should be performed to definitively diagnose moyamoya disease [4,5].

While antiplatelet medications, such as aspirin, are frequently administered to prevent clots, surgery is typically advised instead. There have been various surgeries established for the condition, but the direct method STA-MCA and the in-direct procedures EDAS, EMS, and many burr holes are currently the most popular. The preferred course of treatment is a combined revascularization operation that combines an in-direct technique and a direct Superficial Temporal Artery (STA) to Middle Cerebral Artery (MCA) bypass. The method is believed to lessen the hemodynamic stress on the engorged collateral blood arteries, even if its effectiveness, particularly for hemorrhagic disease, is still unknown. In the frontal and parietal lobes, many burr holes were employed, and excellent neovascularization was attained [6].

- In order to perform the EDAS Encephaloduroarteriosynangiosis (EDAS) treatment, a scalp artery must be severed over a number of centimeters, and a small temporary aperture must then be made in the skull just below the artery. The bone is then restored after the artery is sutured to a branch of the middle cerebral artery on the surface of the brain.
- The temporalis muscle, located in the temple area of the forehead, is dissected and inserted onto the surface of the brain during the Encephalomyosynangiosis (EMS) surgery.
- To allow for the growth of new vessels into the brain from the scalp, many tiny holes (also known as burr holes) are drilled into the skull during the multiple burr holes operation.

It is unknown how this illness develops naturally. When straight bypass is done, the long-term outlook for patients with moyamoya appears to be favorable. The straight STA-MCA procedure immediately results in enhanced blood flow. It is extremely crucial to treat this illness right away since, even with treatment, a massive stroke or haemorrhage may leave the patient with lifelong loss of function.

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