Moyamoya

Definition
Moyamoya originates from a Japanese term, which means “puff of smoke” 1. Moyamoya disease is the progressive occlusion at the termination of the internal carotid artery 2. Stenosis and occlusion of these arteries triggers the development of collateral network of capillaries at the base of the brain 3.

Epidemiology
Incidence of Moyamoya is higher in East Asia area, such as Japan and Korea 4. The annual incidence of Moyamoya in Japan is 0.35 per 100,000 population 3. Females have a higher incidence than males with a ratio of 1.7:1 4. Moyamoya occurs in all age groups but has two peaks with the first peak below age ten and the second peak between 30s and 40s 4.

Pathophysiology
Moyamoya is the progressive stenosis or occlusion of the terminal internal carotid artery 5. The stenosis or occlusion starts in the intracranial internal carotid artery and progresses to the proximal branches of the anterior, or middle cerebral artery 6. Ischemia resulting from the stenosis leads to develop of new and abnormal vascular network around the obstructed area 7. In the cerebral angiogram, the new vessel network or Moyamoya vessels look like a puff of cigarette smoke 8.

Moyamoya with unknown cause is called Moyamoya disease 9. Moyamoya syndrome is used when radiographic features are similar to Moyamoya disease but patients have coexisting medical conditions such as Down syndrome, sickle-cell disease, neurofibromatosis, chronic basilar meningitis, autoimmune mechanism, radiation therapy, and Graves' disease that contribute to the vascular pathology 10.

Manifestations
Moyamoya disease may be asymptomatic 2. Manifestations of Moyamoya differ between children and adult patients. In children, transient ischemic attack (TIA) or cerebral infarct is common. In adults, most patients present with intracranial hemorrhage (intracerebral, intraventricular, or subarachnoid hemorrhage), TIA, or cerebral infarct 3. Other presentations include focal neurological deficits, seizures, headache, choreiform movement, and cognitive deficits 11.

Signs and symptoms of Moyamoya can be provoked by hyperventilation caused by crying, exercise, coughing, straining, or playing a wind instrument 10. During hyperventilation, serum carbon dioxide decreases and causes vasoconstriction. Vasoconstriction further decreases the cerebral blood flow on the area which already has reduced perfusion and aggravates the manifestations of Moyamoya such as dysarthria, aphasia, hemiparesis, or seizure 10.

Diagnostic Tests
Fukui et al. 4 have established the diagnostic criteria for Moyamoya, which include 1) stenosis or occlusion at the terminal portion of internal carotid artery and/or at the proximal portion of the anterior and/or the middle cerebral arteries; 2) abnormal vascular networks in the vicinity of the occlusive or stenotic lesions in the arterial phase; 3) these findings should present bilaterally.

Radiodiagnostic tests such as computer tomography, magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA) have been used to diagnose Moyamoya disease.

Computer tomography is able to assess tumor, hydrocephalus, cerebral infract, or intracranial hemorrhage associated with moyamoya disease 6,11.

Magnetic resonance imaging is able to reveal cerebral infarct and MRA is able to detect the cerebral artery stenosis and collateral vessel formation 11. MRI and MRA are non-invasive methods that are safer and easier than conventional angiography and are useful in identifying any hemorrhage and ischemic event 3. However, compared to a cerebral angiogram, MRA is unable to detect occlusion of the smaller blood vessels 6,10. It also tends to overestimate the degree of arterial occlusion and underestimate the collateral vessels 12.

Cerebral angiogram is still classified as the gold standard for diagnosis of Moyamoya disease 3. Cerebral angiogram is able to assess the location of stenosis and development of collateral circulation 3. Aneurysms or AVMs that are associated with Moyamoya disease can also be detected by conventional angiography 13. Images of the external and internal carotid artery during cerebral angiogram are useful for planning treatment for patients with Moyamoya disease 11.
Treatment Options

No effective medical treatment for Moyamoya disease has been identified 10. The goal of medical management for Moyamoya is focused on the use of anticoagulation to prevent thrombosis and subsequent stroke. Calcium channel blockers such as Nifedipine, nimodipine, flunarizine, and verapamil have been used in Moyamoya disease. The vasodilation effect of the calcium channel blockers is believed to be able to slow the progress of arterial stenosis, promote collateral blood flow and prevent ischemic episodes 14.

Surgical intervention has proven to be effective therapy for Moyamoya 3. Surgical treatments include direct bypass, indirect bypass surgery, or combination of direct and indirect bypass. The goal of surgery is to revascularize and restore perfusion to the stenotic and oxygen deprived area 15. This is to reduce the risk of future ischemia and stroke 16,17.

Direct bypass is connecting the superficial temporal artery with a branch of the middle cerebral artery (STA-MCA) 17. STA-MCA bypass has demonstrated effective in reducing the incidence of recurrent ischemic events 15,17. Direct bypass provides immediate results but may be difficult to perform on the pediatric population because of smaller blood vessels 16. It also requires temporary occlusion of the middle cerebral artery during the procedure and may pose a risk of ischemia to that area 16.

Indirect bypass is creating new circulation by grafting tissue with blood supplies to the ischemic area. There are different types of indirect bypass: encephaloduroarteriosynangiosis is moving the superficial temporal artery under the dura mater close to the cerebrum, this procedure may promote angiogenesis and reduce ischemic attacks 15. Encephalomyosynangiosis is placing the temporalis muscle supply by the deep temporal artery on the surface of the brain on top of the pia mater. Over time collateral vessels are form between the muscle and the ischemic brain tissues 15. Pial synangiosis involves suturing the superficial temporal artery to the pia mater 10,16,17,19. Other surgical interventions include encephaloduroarteriomyosynangiosis, omental transplant, and multiple burr holes with encephalodurosynangiosis 16. It may take months for the collateral artery to develop in the indirect bypass strategies. Patient may develop stroke during this period of time 18. Indirect bypass can be used on patients when no acceptable donor or recipient artery for direct bypass is found. It is less invasive, and does not require temporary clamping of large artery during surgery 15.

Nursing Implication

Hyperventilation from playing a wind instrument 3, anxiety, or crying from pain may result in lowering of PaCO2. Decreased serum carbon dioxide concentration, results in cerebral vasoconstriction. Vasoconstriction worsens the already hypo-perfused area and may result in cerebral ischemia 1, which may lead to subsequent ischemia and stroke 13. Preparation of children pre-procedure is to avoid anxiety and providing adequate pain control post-operatively reduces risk of ischemia 20. Avoid intramuscular injection because it is a source of pain 20.

Oversedation related to overuse of opioid can be devastating. It is because hypoventilation induces hypercarbia. Hypercarbia causes dilation of the normal blood vessels and has no effects on the Moyamoya vessels. This produce a reverse steal phenomenon which redirects blood from the Moyamoya vessels to the normal blood vessels and further reduce blood flow in the disease area 20.

Reference


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