

Quiz 48
Cerebrovascular Atherosclerotic
Disease

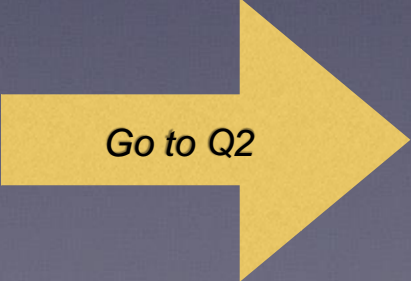
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This quiz is being published on behalf of the Education
Committee of the SNACC.

Start

1. 49 Y/F with a past history of stroke 2 years ago(left arm weakness) and another TIA 1 year ago. Now she presents with weakness and has some cognitive impairment with difficulty in memory and slurred speech. Which of the following is false regarding cerebrovascular occlusive disease?

- A. ICAS(Internal Carotid artery stenosis) usually occurs in middle aged population who present with repeated strokes and cognitive dysfunction.
- B. There can be two types MoyaMoya disease and non-MoyaMoya atherosclerotic internal carotid artery disease.
- C. Extensive collaterals are present in the MoyaMoya group giving a puff of smoke appearance.
- D. Hemorrhagic stroke is unlikely in Moyamoya disease.



Go to Q2

A. ICAS (Internal Carotid artery stenosis) usually occurs in middle aged population who present with repeated strokes and cognitive dysfunction

This is true.

Patients with ICAS were more often young, female, and had Metabolic Syndrome, whereas patients with ECAS (external carotid artery stenosis) were more closely associated with cigarette smoking and hyperlipidemia. ICAS is a more important cause of stroke than ECAS in Asian populations. MCA atherosclerosis was the most important cause of strokes associated with large artery disease. However, proximal ICA was the second most common.

Stroke. 2012 Dec;43(12):3313-8. doi: 10.1161/STROKEAHA.112.658500. Epub 2012 Nov 15.

Risk factors and stroke mechanisms in atherosclerotic stroke: intracranial compared with extracranial and anterior compared with posterior circulation disease.

Kim JS¹, Nah HW, Park SM, Kim SK, Cho KH, Lee J, Lee YS, Kim J, Ha SW, Kim EG, Kim DE, Kang DW, Kwon SU, Yu KH, Lee BC.

Incorrect
Try again

B. They can be two types MoyaMoya disease and non-MoyaMoya internal carotid artery disease.

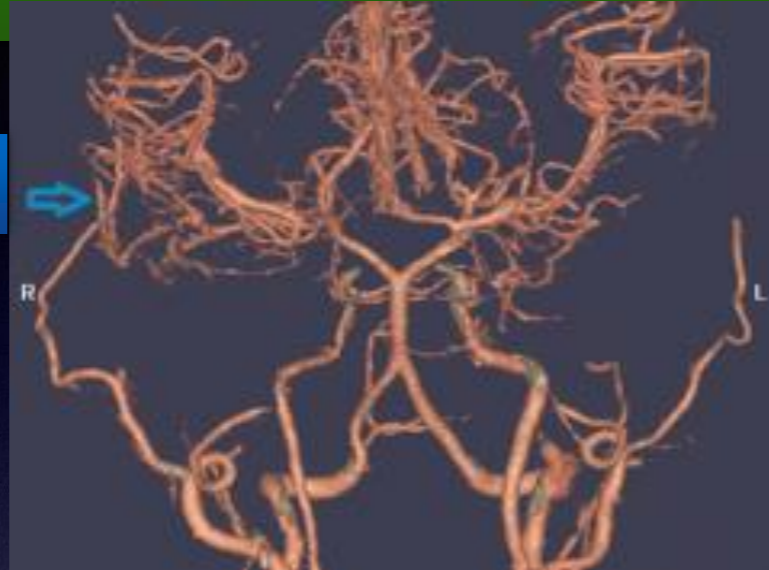
Moyamoya is a disorder where there is progressive spontaneous occlusion of the distal portion of the internal carotid artery which is usually bilateral. This results in extensive hazy collateralization of blood vessels. RNF gene polymorphism seems to be a susceptibility factor.

It is often difficult to differentiate moyamoya disease from non-MoyaMoya intracranial cerebrovascular occlusive disease in adult patients. Both present with similar constellation of symptoms. However the puff of smoke appearance due to collateralization may not be present.

Incorrect
Try again

C. Extensive collaterals are present in the MoyaMoya group giving a puff of smoke appearance.

Collaterals



This is True and is more common in the MoyaMoya group.
this disease was named MD due to network of vessels appearing as a “puff of smoke” on conventional angiography.
It consists of leptomeningeal vessels and lenticulostriate arteries which give rise to the characteristic puff of smoke appearance.

Incorrect
Try again

D. Hemorrhagic stroke is unlikely in Moyamoya disease.



This is the correct answer.
The above statement is false.

Hemorrhagic stroke can occur in this disease. The vascular dilation occurring in the thin walled collaterals can lead to micro-aneurysmal dilation and rupture. hemorrhagic stroke can also occur in the postoperative period when blood pressure control is inadequate.

Robert J.W.Ryan etal; Surg Neurol Int. 2012; 3: 72.
Hemorrhage and risk of further hemorrhagic strokes following cerebral revascularization in Moyamoya disease: A review of the literature

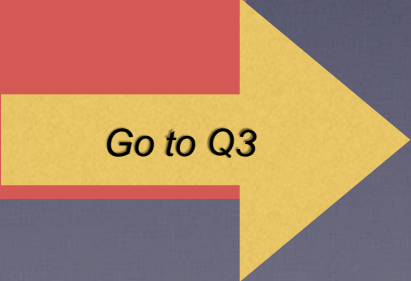
Back to Q

Next Q

2. The following are associated with development of Moya Moya syndrome except

- A. Downs Syndrome
- B. Von Recklinghausen's Disease
- C. Cranial Irradiation
- D. Craniosynostosis

Go to Q3



A. Down's Syndrome

When Moyamoya-like vasculopathy develops in association with systemic diseases and conditions, it is termed Moyamoya syndrome. While Moyamoya disease is idiopathic, the syndrome is secondary.

Moyamoya Associations

Neurofibromatosis -1

Downs syndrome

Thyroid Disease

Cranial Irradiation

Sickle cell anemia

Rare Diseases

SLE

Turners

Olivers syndrome

Ji Hoon Phi,, et al; Moyamoya Syndrome: A Window of Moyamoya Disease.

J Korean Neurosurg Soc. 2015 Jun; 57(6): 408–414

Incorrect
Try again

B. Von Recklinghausen's Disease

NF-1 (neurofibromatosis type 1), also known as von-Recklinghausen disease, is a syndrome caused by a germline loss of one NF1 gene allele. NF-1 is an autosomal dominant disorder. NF-1 involves multiple organs, including the brain, peripheral nerves, eyes, skin, and bones.

In the brain, it can manifest as optic glioma, macrocephaly, learning and cognitive disabilities and cerebral vasculopathy which includes stenosis/occlusion and aneurysmal dilatation.

Ji Hoon Phi, et al; Moyamoya Syndrome: A Window of Moyamoya Disease.
J Korean Neurosurg Soc. 2015 Jun; 57(6): 408–414

Incorrect
Try again

C. Cranial Irradiation

This is True.

The risk of developing Moyamoya syndrome is highest for patients who are irradiated for tumors typically located around the circle of Willis where the moyamoya pathognomonic vascular changes occur, such as optic glioma, craniopharyngioma, and germ cell tumors

Keene DL, etal. Vascular complications of cranial radiation. Childs Nerv Syst. 2006;22:547–555.

Incorrect
Try again

D. Craniosynostosis



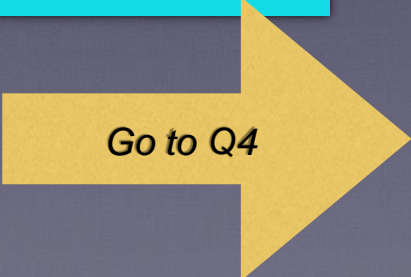
Craniosynostosis has not been associated with Moyamoya syndrome and hence this is the correct answer.
Craniosynostosis is a condition where there is premature closing of skull sutures.

Back to Q

Next Q

3. The following are true regarding cerebrovascular reactivity **EXCEPT**

- A. Patients with an increased oxygen extraction fraction (OEF) measured by positron emission tomography (PET) have an increased risk of recurrent stroke.
- B. Patients with increase in flow velocity during apnea testing assessed by transcranial Doppler sonography have an increased risk of recurrent stroke
- C. Blood oxygen level dependent (BOLD) functional MRI can detect areas of low cerebral reactivity
- D. Acetazolamide can be used as a provocative test for detection of areas of low cerebral vascular reactivity



Go to Q4

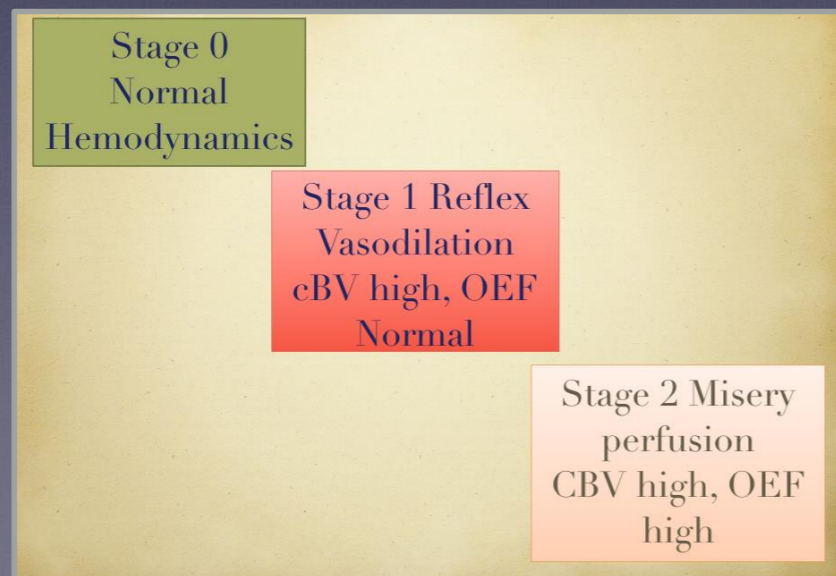
A. Patients with an increased oxygen extraction fraction (OEF) measured by positron emission tomography (PET) have an increased risk of recurrent stroke.

This is True.

PET scans can determine microvascular cortical perfusion, and oxygen extraction fraction (OEF). Patients with an increased oxygen extraction fraction (OEF) measured by positron emission tomography (PET) and abnormal cerebrovascular reactivity measured by other techniques such as Transcranial Doppler can have an increased risk of recurrent stroke.

The disadvantages of PET is that it is not widely available and the examination carries a radiation load.

Stages of Cerebrovascular Hemodynamic Failure



Incorrect
Try again



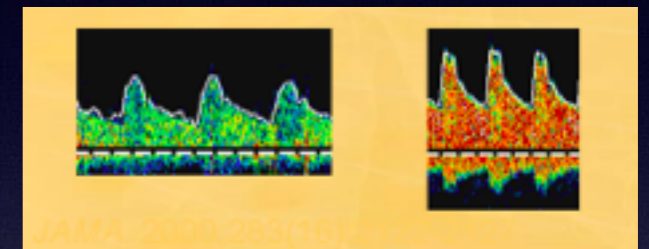
B. Patients with increase in flow velocity during apnea testing assessed by transcranial Doppler sonography have an increased risk of recurrent stroke

This is false.

Patients with NO CHANGE or DECREASE in flow velocity during apnea testing assessed by transcranial Doppler sonography have an increased risk of recurrent stroke

Provocative apnea testing is done using Transcranial Doppler(TCD) to determine Cerebrovascular reactivity. Apnea causes rise in CO₂ tension leading to increase in mean flow velocity and pulsatility index in the TCD.

Breath holding for 30 seconds should increase the velocity by approximately 20%. If this does not happen, one can determine that cerebrovascular reactivity is poor thereby leading to increased risk of stroke.



baseline
TCD

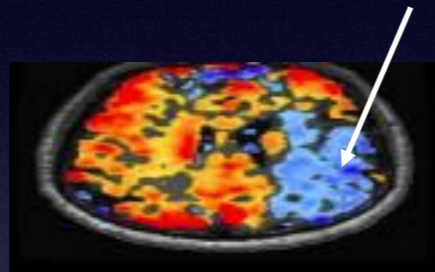
TCD after
apnea

Back to Q

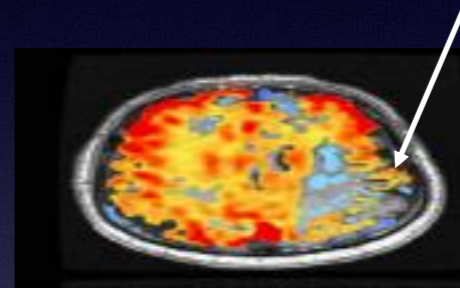
Next Q

C. Blood oxygen level dependent (BOLD) functional MRI can detect areas of low cerebral reactivity

Area of low CVR



Improved after bypass



A recent study by Mandell et al determined Cerebrovascular reactivity by using BOLD (Blood oxygen level dependent MR imaging) and CO₂ as a vasodilatory stimulus.. Their conclusion was that preoperative CO₂ BOLD CVR predicts the hemodynamic effect of ECIC bypass surgery in patients with intracranial steno-occlusive disease.

D M Mandell et al ;Quantitative measurement of Cerebrovascular Reactivity by Blood Oxygen Level-Dependent MR Imaging in Patients with Intracranial Stenosis: Preoperative Cerebrovascular Reactivity Predicts the Effect of Extracranial-Intracranial Bypass Surgery. AJNR Am J Neuroradiol 32:721–27 Apr 2011;

Incorrect
Try again

D. Acetazolamide can be used as a provocative test for detection of areas of low cerebral vascular reactivity

This is True.


Acetazolamide injection causes vasodilation just like CO₂ and can be used as a provocative test in combination with MR imaging or SPECT scan(single-photon emission computerized tomography).

Areas of low cerebrovascular reactivity improve in images taken after an Extracranial-intracranial bypass.

Incorrect
Try again

4. Which of the following is **TRUE** regarding anesthesia management for a patient undergoing an extracranial-intracranial bypass (superficial temporal artery to middle cerebral artery)?

- A. Total intravenous anesthesia has found to be more beneficial in outcome studies than using inhalation agents.
- B. It is important to keep the blood pressure on the lower side before cross clamping and higher after anastomosis
- C. Patients who have pressure dependent symptoms preoperatively have a higher chance of stroke after surgery.
- D. Hypercapnia during surgery can cause postoperative stroke but hypocapnia is safe.



Go to Q5

A. Total intravenous anesthesia has found to be more beneficial in outcome studies than using inhalation agents.

This is False.

Studies have shown no difference whether TIVA or inhalation anesthesia was used for these procedures. In a study assessing the perioperative course of 216 patients undergoing revascularization surgery, there were no differences in patient outcome among different anesthetic techniques (inhaled, IV, and balanced). The severity of disease and surgical procedures were the only determinants of neurological deterioration.



Sakamoto T, Kawaguchi M, et al;
Risk factors for neurologic deterioration after
revascularization surgery in patients with moyamoya disease.
Anesth Analg 1997;85:1060-5

Incorrect
Try again

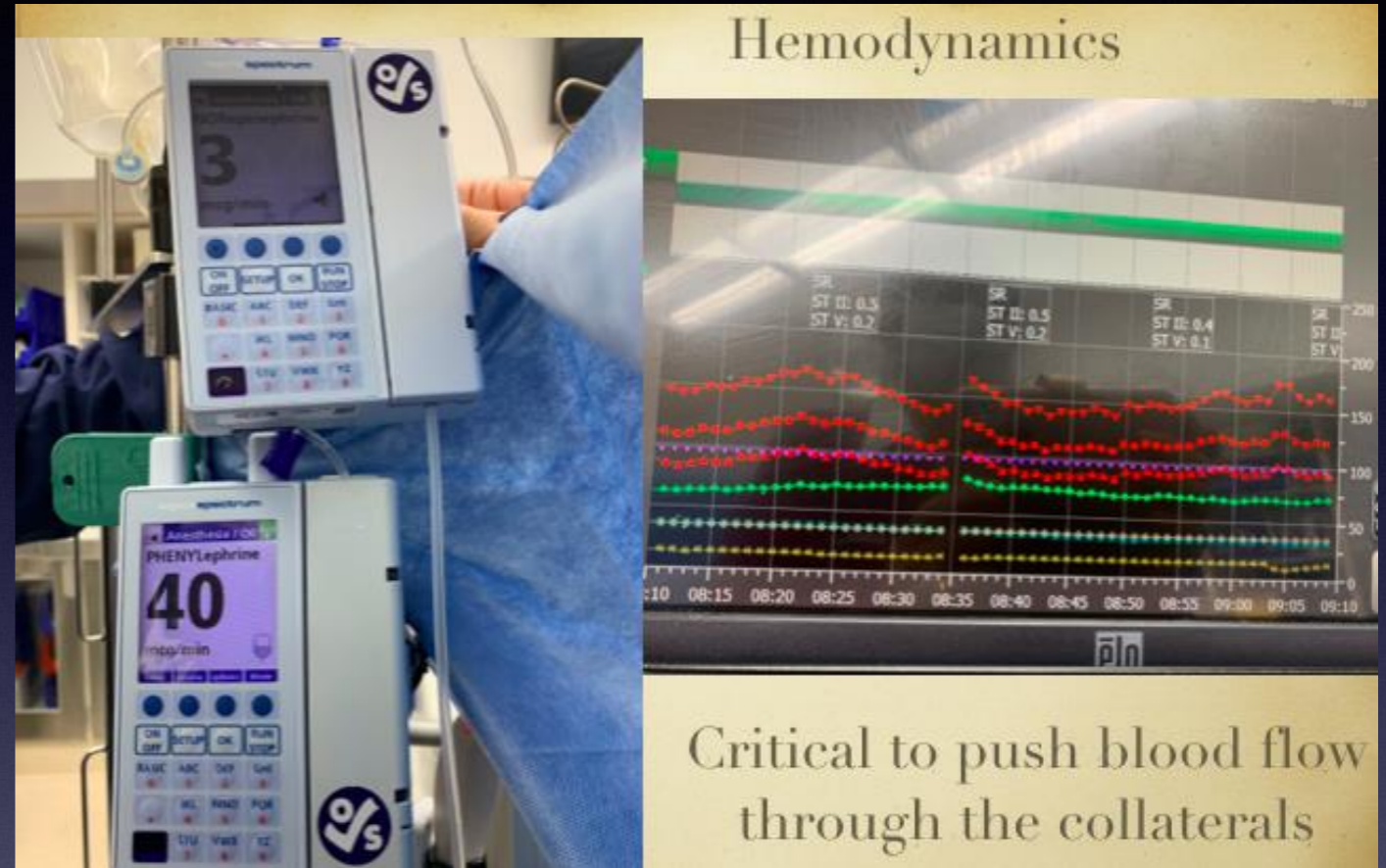
It is important to keep the blood pressure on the lower side before cross clamping and higher after anastomosis

This is false.

Hypotension is poorly tolerated particularly during the clamping phase and

could lead to cerebral ischemia. The general recommendation to keep the blood pressure 10% to 20% above baseline during clamping to push blood flow through the collaterals.

After anastomosis the blood pressure should be kept normotensive to prevent bleeding at the anastomotic sites and to prevent cerebral edema due to hyperperfusion.



Incorrect
Try again

Patients who have pressure dependent symptoms preoperatively have a higher chance of stroke after surgery.



This is True.

Patients who have neurologic symptoms if the blood pressure is dropped are at higher risk for postoperative stroke as evidenced by the study by . It is important to identify this group of patients preoperatively and keep their blood pressure extremely stable.

Back to Q

Next Q

D. Hypercapnia during surgery can cause postoperative stroke but hypocapnia is safe.

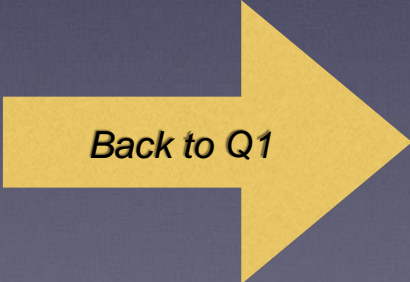
Normocapnic Normoxemia is the goal. Both Hyper and hypocapnia can be deleterious. Hyperventilation-induced hypocapnia causes cerebral vasoconstriction and can lead to cerebral ischemia especially in areas which have poor Cerebral Blood Flow. Hypercapnia can also have undesirable effects. The collateral network of vessels in patients with moyamoya are already maximally vasodilated. Hypercapnia can cause an “intracerebral steal” effect when other normal vessels vasodilate leading to cerebral ischemia and poor outcomes.



Incorrect
Try again

5. Postoperative complications after bypass surgery in Moyamoya include the following **except-**

- A. Communicating Hydrocephalus
- B. Ischemic Stroke
- C. Cerebral edema
- D. Hemorrhagic Stroke



Back to Q1



A. Communicating Hydrocephalus

This is not a complication.
Hence this is the correct answer for this question.

Back to Q

Next Q

B. Ischemic Stroke

Ischemic stroke is a complication.

Postoperative infarction is closely related to the hemodynamic instability characteristic of MMD. Many studies have found that several factors, such as hypercapnia, hypocapnia, hypotension and inadequate hematocrit, increase the risk of postoperative ischemia In 1997,

Sato et al. have identified 3 specific risk factors, including the presence of a preoperative low-density area, intraoperative urinary output, and reduced hematocrit In addition, early graft occlusion in an unblocked bypass can lead to cerebral ischemia. Acute preoperative infarcts and poor cerebrovascular reserve are independent risk factors for severe ischemic complications following direct extracranial-intracranial bypass in MMD

Ref: Jinlu Yu et al; Progress on Complications of Direct Bypass for Moyamoya Disease. Int J Med Sci. 2016; 13(8): 578–587.

Incorrect
Try again

C. Cerebral edema

Postoperative hyperperfusion can cause excessive proliferation and edema of endothelial and smooth cells found in the arteriolar walls causing cerebral edema just like in CEA(Carotid endarterectomy).

Ref:

1. Progress on Complications of Direct Bypass for Moyamoya Disease Jinlu Yu, Lei Shi, Yunbao Guo, Baofeng Xu, and Kan Xu Int J Med Sci. 2016; 13(8): 578-587.
2. Haruto Uchino, MD et al; Predictors and Clinical Features of Postoperative Hyperperfusion after Surgical Revascularization for Moyamoya Disease -A Serial Single Photon Emission CT/ Positron Emission Tomography Study

Incorrect
Try again

D. Hemorrhagic Stroke(HS)

There is risk of hemorrhage after treatment with either direct or indirect bypass both in the immediate as well as long-term future. Persistence of microaneurysms and damaged vessel walls, loss of cerebral autoregulation, and underlying microangiopathy with microbleeds are all possible etiologies.

The most serious consequence of HS is intracranial hemorrhage. HS-induced hemorrhage includes intracranial hematoma and subarachnoid hemorrhage. Most postoperative cerebral hemorrhages occurring after MMD direct bypass are caused by excessive perfusion, while some are caused by hemodynamic changes. However, contralateral cerebral hemorrhage is not considered to be caused by hyperperfusion. This type of hemorrhage is speculated to be associated with perioperative hemodynamic changes and contralateral vascular rupture hemorrhage induced by anesthesia.

Jinlu Yu et al; Progress on Complications of Direct Bypass for Moyamoya Disease.
Int J Med Sci. 2016; 13(8): 578–587. Published online 2016 Jul 5.

Back to Q 1

End of Set

Incorrect
Try again