**Myringotomy Tube Disaster – Internal Carotid Bypass in Pediatric Patient With Moyamoya Disease and Poor Circle of Willis Integrity**

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**Goals**

1. Review of aberrant anatomy that can result in significant blood loss during myringotomy tube placement.
2. Proper management following puncture of the internal carotid artery during myringotomy tube placement (AKA Crisis Management in the OR)
3. Anesthetic management of pediatric patient requiring cerebral angiogram in the interventional neuroradiology suite – asleep-awake-asleep vs. general anesthesia
4. Anesthetic management of patients that require SSEP/MEP monitoring, and effectiveness of balloon test occlusion of the internal carotid artery
5. Management of artery to artery bypass and the implications in a patient with Moyamoya disease

**Description**

An 11-year old boy with a history of Moyamoya Disease underwent bilateral myringotomy tube placement at an outpatient surgery center. During the procedure, the otolaryngologist inadvertently punctured a large vessel behind the right tympanum, and the anesthesiologist noted approximately 700cc of blood loss. After packing the patient's ear, the bleeding stopped and the patient was sent home. One week later, after a violent cough, his mother noted hemorrhaging from the ear, and he presented emergently to the interventional neuroradiology suite for angiography. Blood was transfused for a hematocrit of 19, and the angiogram confirmed a vascular insult to the right internal carotid artery. A balloon occlusion was performed to determine the integrity of the Circle of Willis, but within one minute of the occlusion, the patient was noted to lose both SSEP and MEP signals on the left side of the body. He was immediately transported to the OR, where a craniotomy was performed to allow for an internal carotid to middle cerebral artery bypass using a radial arterial graft. This case highlights the dangers of aberrant vascular anatomy during myringotomy tube placement, the importance of SSEP and MEP monitoring, and the effects of Moyamoya Disease on arterial integrity.

**Case history:**

An 11 year old, 40 kg male, presents to you for angiography in the neurointerventional

radiology suite. He is currently receiving a blood transfusion through a 20G PIV.

Questions:

What further information would you like to have? Are there any special considerations you want to take into account based on his history of Moyamoya disease? What is Moyamoya disease?

**Case history (continued):**
The patient is deaf in the left ear and is now unable to hear on the right because of the hemorrhage. Aside from his Moyamoya disease, and mild obesity, the patient is otherwise healthy. He was noted to have 700cc of blood loss from bilateral myringotomy tube placement done at an outpatient surgery center two weeks prior to his arrival today. Moyamoya is named after "a puff of smoke" in Japanese, and is named so because of collateral vessels that form in an attempt to improve blood flow through an obstructed or non-patent circle of Willis. The disease mainly affects the internal carotid arteries, and the intracranial vessesls (typically the MCA/ACA), but can extend to extracranial vessels as well. These collateral vessels are weak, have poor flow, and are prone to thrombosis or injury, which can result in aneurysmal changes or hemorrhage. Unfortunately, this disease is progressive, lifelong, and not amenable to medical management. If symptomatic enough, the only treatment that has been effective is cerebral revascularazation via a bypass graft.

**Questions:**
Is there any specific anesthetic technique that would prove to be advantageous for this case? Do we need to put this patient to sleep? What would you want to use for maintenance?

**Case history (continued):**
Ideally, this patient would best be done with sedation so that the patient could follow commands and move his legs during occlusion of the affected carotid artery. Multiple agents such as dexmedetomidine, or midazolam with narcotics could be utilized. Unfortunately, in this patient's case, there would be no way for us to ask him to follow commands, as he is unable to hear in either ear. While writing or the use of flash cards could be attempted, the sedation required for placing large bore catheters in the groin may preclude the ability to respond in a timely manner for commands. Finally, should a test occlusion of the carotid artery result in severe neurological changes, this could become a dangerous situation that would put the patient at more risk.

The combination of his underlying and iatrogenic deafness, young age, and mild obesity, the decision was made to proceed with general anesthesia. Of course, upon induction, while small amounts of paralytics could be utilized to secure an endotracheal tube, it is preferable to use a smaller dose of a nondepolarizing neuromuscular blocker, followed by reversal, a dose of succinylcholine, or a method via which a paralytic is not required.

Due to the monitoring of SSEP's and MEP's, it would be advisable to use a total intravenous anesthetic (TIVA). Much debate can occur between neuro-monitoring technologists and

anesthesiologists, however, what is imperative is the need for good signals to be interpreted by the technologist. Caveats for SSEP and MEP monitoring are that volatile agents are the most suppressive, followed closesly by N2O. Propofol, dexmedetomidine, and narcotics are all moderately suppressive of these signals, and agents such as ketamine, etomidate, and lidocaine all the the least effects. SSEP's are more sensitive than MEP's and are more sensitive to all of the above anesthetic medications.

The patient was intubated using a combination of lidocaine, propofol, narcotic, and sevoflurane. After endotracheal intubation, the volatile anesthetic was discontinued, and the patient was maintained on an infusion of propofol, dexmedetomidine, and fentanyl.

Angiography confirmed an injury to the right internal carotid artery. The next step by the surgeon was to occlude the internal carotid and determine the integrity of the Circle of Willis in providing adequate collateral flow from the contralateral side.

**Questions:**
What changes will you look for in the MEP tracings? What about your anesthetic can change the evoked potentials? How can you confirm that if MEP changes occur, that it is a result of balloon occlusion, and not your anesthetic?

In this case, the MEP tracings would go flat on the left side of the body (remember that the contralateral side is effected). Anesthetics can have a tremendous impact on the MEP tracing, and need to be carefully monitored. Temperature, hematocrit, and blood pressure are the most important, followed by volume status and cardiac output.

Of course, if a bolus of an anesthetic agents such as propofol is administered by the anesthesiologist, this can immediately dampen or even eliminate motor evoked potentials. In this case, following the balloon occlusion of the right internal carotid artery, the MEP's on the left upper and lower extremity disappeared entirely. Upon release of the balloon, the MEP tracings reappeared within 30 seconds. This was done three times to ensure that this was not as a result of anesthetic changes.

The most important element in this is to have good communication between the surgeon, the anesthesiologist, and the technologist that is interpreting the tracings. Having a good baseline of tracings will help to determine if the loss of signal is due to a physiological error, a surgical intervention, or a change in the anesthetic. Drastic drops are going to be more commonly related to a surgical complication such as a misplaced screw in the spine, a catastrophic amount of blood loss, or a bolus by the anesthesiologist. More slow loss of signal can be indicative of suboptimal physiological parameters, whether it is a slow amount of blood loss, a drift in the core temperature, or a gradual drop in the blood pressure.

In this case, the loss of MEP signals was clearly and temporally related to the occlusion of the carotid artery.

At this point, the decision was made to proceed to the operating room to perform an internal carotid to middle cerebral arterial bypass, using a radial artery graft.

**Questions:**
How will you handle the consent process with the family? What further access would you like to have for this case? Is a central line warranted? If so, where would you place it? What implications does

Moyamoya disease have for arterial catheterization? What blood products would you want to have on hand?

While consent for anesthesia is generally implied or understood when surgical interventions are being performed, it is important to communicate with the family if major changes in the anesthetic plan are anticipated. If a fellow, resident, or CRNA is available, and it is an appropriate time to speak with the family, then this would be the best scenario. In the case that you are administering the anesthetic on your own, having a telephone conversation with the family would be important.

While a CVC is not required in this case, the combination of pre-existing anemia, possibility for prolonged surgery, and need for infusions would suggest that placement of a CVC would be advantageous for this patient. There has been some discussion with the occlusion of the IJ by the central line causing cerebral venous drainage issues, however, an appropriate sized cannula should not cause any clinically significant changes in this drainage.

Moyamoya does carry the theoretical risk of extracranial arterial involvement, however, this is uncommon, and would be extremely unlikely in an 11-year old patient. Arterial catheterization was easily performed in this patient using ultrasound guidance on the right side. The surgeon harvested the left radial artery for grafting, after ensuring adequate flow to the hand was achieved with only the ulnar artery. The left radial artery was clamped for several minutes prior to removal.

The right carotid artery was successfully grafted and anastomosed to the middle cerebral artery. The right internal carotid was then tied off above the graft, and the patient went on to make a successful recovery, with the hearing eventually returning to normal in his right ear.

**Discussion**

This case brings up several unique challenges. First, the challenge of diagnosing this patient while asleep required us to come up with a creative management plan to help accurately diagnose the integrity of the Circle of Willis.

The need for MEP monitoring under general anesthesia, and the implications of a failed occlusion study made it imperative to understand the results of the study. In this case, a major intracranial procedure was required following the results of a failed balloon occlusion of the internal carotid artery. Had the test been negative, a complete occlusion of the right internal carotid artery could have been performed, removing the need for a bypass graft on that side.

Ultimately, this all stemmed from the placement of a myringotomy tube, something that was not brought up during the case discussion. The aberrant location of the internal carotid artery sometimes places the vessel directly behind the tympanum. In the case of myringotomy tube placement, this aberrancy can result in catastrophic consequences, as this patient experienced. While not common, it is a unique issue that should be understood by both the anesthesiologist and the ENT.

In this patient's case, the final outcome was positive, in that his hearing returned in the right ear, however, he went through quite a long process. The complicating factor in this patient was the existence of his Moyamoya disease, which necessitated a bypass of his internal carotid

artery, whereas a patient with an intact Circle of Willis would not have needed further intervention.

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