# **Anesthetic Implications of** Moyamoya disease: A case study

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## INTRODUCTION

- Moyamoya disease (MMD) is a rare condition with significant anesthetic implications and few guidelines<sup>1,2</sup>
- The condition is characterized by progressive narrowing and stenosis of the distal internal carotid artery (ICA) and proximal middle cerebral arteries (MCA), anterior cerebral arteries (ACA) and less commonly posterior cerebral arteries (PCA). <sup>1</sup>
- Most evidence exists as reviews or retrospective studies based on revascularization surgery specifically for MMD
- A knowledge gap exists due to unfamiliarity of MMD and no standard guidelines for management exist

## **PURPOSE STATEMENT**

• This case study educates anesthesia providers about MMD and provides recommendations for management during the perioperative period

## CASE SUMMARY

- 34-year-old female, ASA physical status 3
- PMH CVA r/t MMD, chronic mastoiditis and recurrent otitis media
- Presented for L mastoidectomy and ear tube placement
- Preoperative arterial line placed
- General endotracheal anesthesia, second large-bore IV, cerebral oximetry
- Maintenance of anesthesia with volatile anesthetic
- Blood pressure augmentation with phenylephrine infusion and maintained near baseline
- Pain management with IV boluses of hydromorphone
- Extubated deep per surgeon request
- Transferred to post-anesthesia care unit (PACU) with no new neurologic symptoms and level of consciousness to baseline

## METHODS

- PubMed and EMBASE searched with the terms "moyamoya" disease" AND "anesthesia" and "moyamoya disease" AND "perioperative"
- Results narrowed to the last 5 years and to exclude pregnancy to yielded 36 and 92 respectively
- Results included retrospective studies, case studies, and reviews
- UpToDate searched with the term "moyamoya disease" to retrieve background information

cerebrovascular disease with significant anesthetic implications and limited evidence exists to guide anesthesia providers for

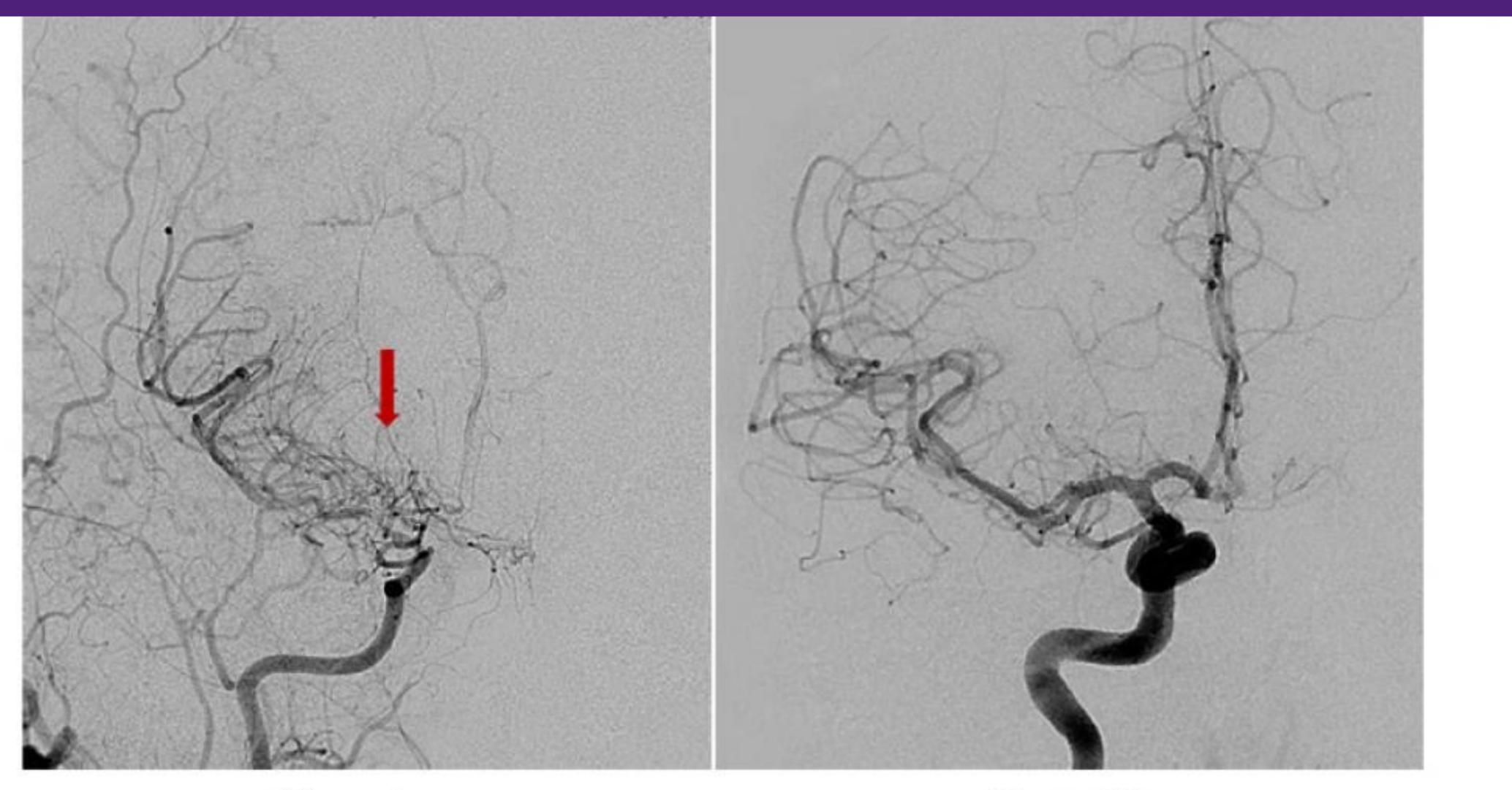


Figure 1a

**Figure 1a.** AP angiogram imaging of moyamoya disease.<sup>6</sup> Moyamoya translates to "hazy puff of smoke" in Japanese, which are created by small collateral vessels and can be seen in this image.<sup>1,6</sup> Figure 1b. A normal angiogram for comparison.<sup>6</sup>



# Moyamoya disease is a rare perioperative care

Figure 1b



## **SUPPORTING EVIDENCE**

- MAP was the most commonly suggested BP measurement and adjusting goals to patient's baseline blood pressure<sup>2,3,4,5</sup>
- Keeping MAP between 85 and 90% of baseline was also discussed<sup>3,5</sup>
- Blood pressure augmentation preference varied greatly from crystalloid/colloid infusion and choice of vasopressors<sup>2,3,4,5</sup>
- Carbon dioxide management is critical due to the effect on cerebral blood flow and intracranial pressure (ICP)<sup>2,3,4,5</sup>
- Maintain fluid status from euvolemic to hypervolemic for revascularization surgery <sup>2,3,5</sup>
- Hypothermia did not show a difference in positive outcomes<sup>3</sup>
- Hyperthermia can increase metabolic demand in the brain<sup>3</sup>
- While no definitive evidence exists, cerebral oximetry could aid in hemodynamic management<sup>3</sup>

## DISCUSSION

- There is no single type of anesthetic technique that favors more positive neurologic outcomes for MMD patients<sup>3,4,5</sup>
- Blood pressure management appears to be one of the critical parts of MMD management and warrants arterial line
- The consensus of zeroing/placing arterial line transducer is at the tragus (Circle of Willis)<sup>2</sup>
- Ways to assess fluid status include urine output and pulse pressure variation (PPV)<sup>3,5</sup>
- Normocarbia was the most commonly suggested parameter for ventilation management
- Normothermia should be maintained

## CASE CRITIQUE

 More balanced anesthetic with gas and remifentanil infusion could have decreased risk of cerebral steal

## **RECOMMENDATIONS FOR PRACTICE**

- Future studies should focus on TIVA vs GA vs combined technique
- Blood pressure should be maintained no lower than 85% of baseline MAP
- A preoperative arterial line for adults should be placed to manage hemodynamics and maintained at the level of the tragus
- Based on lack of evidence, specific vasopressors cannot be recommended
- Fluid goal strategies are patient and procedure driven
- Maintenance of normocarbia can minimize risk of cerebral steal and increased ICP
- Normothermia to hypothermia is safe with strict avoidance of hyperthermia
- Use of cerebral oximetry has potential benefits with low risk and simple application

## CONCLUSIONS

• The limited evidence about MMD and anesthetic management not only justifies this case study but also presents significant challenges related to the disease process and lack of evidence for perioperative management