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Perioperative Management of a Patient With Moyamoya Disease and Pre-eclampsia Undergoing Cesarean Section.

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Abstract Body: Moyamoya disease is characterized by progressive bilateral stenosis of the distal internal carotid arteries and their branches which reduces blood flow to the vessels of the anterior circulation of the brain. Inadequate perfusion promotes the development of an extensive network of collateral vessels which have a characteristic hazy appearance on imaging. Affected individuals are predisposed to ischemic stroke as well as hemorrhagic events involving the fragile collaterals, and pregnancy may exacerbate these risks. Anesthesia for these patients poses significant risks. Multiple perioperative strategies have been reported in the literature, but there are no established guidelines for optimal management.

RR, a 20-year-old G1P0 at 37 weeks, was noted to have progressive hypertension, proteinuria and extremity edema at an outside hospital. She had a history of multiple ischemic strokes and Moyamoya disease, and was five years status post successful bilateral cerebral revascularization procedures with no subsequent ischemic events.

Upon presentation at our facility, RR continued to be hypertensive. Her physical exam was otherwise unremarkable. Laboratory values were within normal limits. Brain MRI and MRA performed upon admission showed areas of remote infarction but confirmed adequate perfusion and no acute intracranial abnormalities.

In view of suspected pre-eclampsia, RR underwent an elective low transverse cesarean section under epidural anesthesia with invasive arterial pressure monitoring. She was hemodynamically stable throughout the procedure and suffered no neurological insults. Her postoperative course was uncomplicated and she was discharged home on post-operative day number three.

The guiding principles in anesthetic management of patients with Moyamoya are maintenance of normotension and normocapnia along with careful monitoring of neurological status. Both hypotension and hypocapnia can precipitate ischemic attacks. Hypercapnia may also result in decreased blood flow in affected areas secondary to a cerebral steal phenomenon. Significant hypertension must be avoided due to increased risk for intracranial hemorrhage.

Neuraxial anesthesia is associated with hypotension as well as anxiety related hypocapnia and hypertension. We considered these risks manageable with careful technique and judged a neuraxial anesthetic a safer choice than general anesthesia. Neuraxial anesthesia allows avoidance of the hypertension that often follows laryngoscopy and intubation, and it facilitates continuous monitoring of neurological status. Epidural anesthesia was considered preferable to spinal anesthesia because the associated hemodynamic changes are generally more gradual and therefore easier to carefully control with pressors and fluids. Anxiolytics were cautiously administered in order to minimize the risk of intraoperative hyperventilation and hypertension related to anxiety.