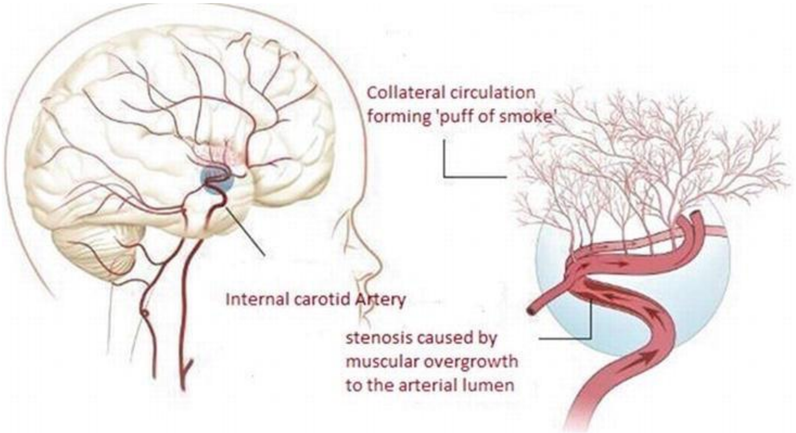
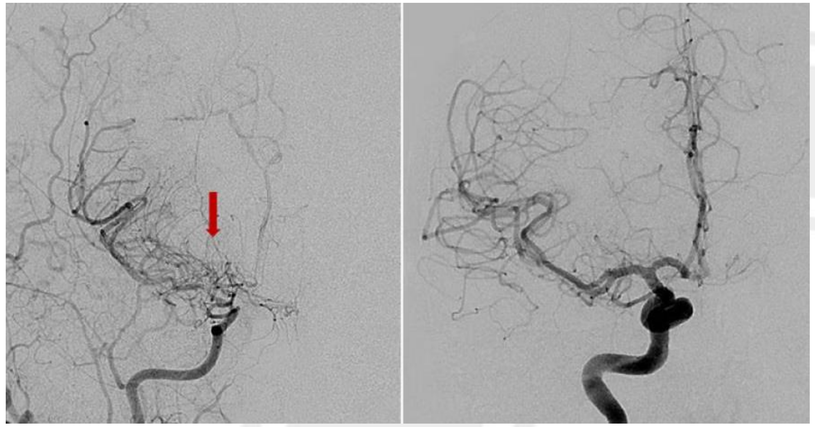
Moyamoya Disease in Pregnancy

Cassie Wernke, DO; Leslie Matthews, MD, PharmD; Teri Gray, MD

Moyamoya Disease Defined:

- Bilateral, chronic progressive occlusion of terminal portion of internal carotid arteries→abnormal collaterals
- Risk of cerebral ischemia and hemorrhage
- 2003 Japan Survey: prevalence and incidence were 6 and 0.54 per 100,000 respectively. (1)
  - M:F 1:1.8
  - 90% of diagnosed <40yo





1. Inayama, Y. et al. (2009) World Neurosurgery 122: 684-691

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Case:

- 35F G2P0010 at 35w3d gestation presented for IOL due to gestational HTN w/ severe features.
- PMH: Moyamoya disease w/ multiple TIAs, PFO s/p repair, gestational diabetes
- Pt was induced, epidural anesthesia was initiated.
- Neurosurgery consulted- No absolute contraindication to vaginal delivery, but risk with maternal valsalva.
- Offered assisted second stage labor vs. primary cesarean delivery.
- Complete cervical dilation but arrest of fetal descent.
  - → primary Cesarean delivery
- Required treatment for severe range blood pressures throughout her course, otherwise underwent uncomplicated C/S and discharged on POD #3.

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Key Takeaways:

- Physiologic changes in pregnancy may worsen symptoms of disease
  - 50% increase in blood volume, hypercoagulability, stress of L&D
- Optimal mode of delivery?
  - C/S- hemodynamic changes with laparotomy and intubation, fluid shifts, bleeding
  - Vaginal w/ epidural- HTN with second stage of labor, hyperventilation w/ pain/anxiety
- Neuraxial anesthesia is not contra-indicated (2, 3)
  - Avoid hypertension, hypotension, hyperventilation (2)
    - CPP=MAP-ICP
  - Further complicated by hypertensive disorders of pregnancy
    - Intracerebral hemorrhage vs. HTN disorder?
    - Aggressive blood pressure control, maintain normovolemia, and limit blood loss
  - Multidisciplinary planning w/ in depth risks/benefits discussion- OBGYN, Neurosurgery, Anesthesiology throughout peri-operative period (3)

2. Komiyama, M. et al. (1998) Neurosurgery 43: 360-368
3. Sato, K. et al. (2015) Journal of Stroke & Cerebrovascular Diseases 24: 921-924

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