

Anesthetic management of a child with Down syndrome and Moyamoya disease for surgical revascularization

Thomas E. Cox, MD

John D. McAllister, MD

Objectives

After discussing this case, the participant should be able:

1. Discuss the pathophysiology of Moyamoya disease as well as the medical and surgical approaches to therapy.
2. Identify the psychosocial and clinical issues relevant to the care of the child with Down syndrome in the perioperative period.
3. Discuss strategies for the anesthetic management of a challenging patient with Moyamoya disease for surgical revascularization.

Stem Case and Key Questions

A 10-year-old, 28 kg male with Moyamoya disease is scheduled for a revascularization procedure (EDAS). His history is significant for Trisomy 21, obstructive sleep apnea, and hypothyroidism. In addition, the child had a stroke 6 months prior to admission and has a mild residual left hemiparesis. Medications include levothyroxine 75mcg qD and aspirin 81mg qD. P 72, BP 90/58, R 22, T 37.0°C.

What is Moyamoya disease?

What are the medical and surgical options available to treat Moyamoya disease?

What additional information would you like prior to this patient's admission for surgery?

The patient receives a preoperative MRI, cerebral angiogram, and CT scan. The diagnosis of Moyamoya disease is confirmed with bilateral carotid artery stenosis (R>L) and evidence of previous bilateral infarcts in the middle cerebral artery distribution. The medical and surgical options are presented to the parents. Consent is obtained to proceed with bilateral revascularization (EDAS).

In general, what are your anesthetic concerns in a patient with a history of cerebral vascular ischemia and/or infarct? Trisomy 21? Developmental delay and/or uncooperative behavior? Obstructive sleep apnea?

What are your options for the premedication and induction of a child with developmental delay and OSA?

On the day of surgery, the child is assessed to be calm and cooperative. No premedication is given and mom is given instruction and allowed to accompany her child to the OR for an inhalational induction. Upon entering the OR, the child becomes uncooperative and refuses to get onto the OR bed.

What are your options now? ... Abort and premedicate? ... IM ketamine? ... Restrain and proceed with an inhalational induction?

Discuss the advantages and disadvantages of each option given this patient's risk factors.

Following an inhalational induction with O₂ / N₂O / Sevoflurane, the monitors are placed and surgical preparation proceeds.

Will you utilize any special monitoring for this surgery?

How will you maintain anesthesia for this surgical revascularization procedure? Is there a decided advantage to one agent(s) and/or technique(s) over another? Regarding CMRO₂ and CBF?

What are your goals for the ventilatory management of this anesthetic?

While the neurosurgeon is dissecting out the superficial temporal artery and galeal strip, the child's BP increases from 110/60 to 160/90.

What is your differential diagnosis for this increase in BP? How will you assess and will you treat?

What are your hemodynamic goals for this case? Are they different than for other intra-cranial procedures?

The surgical revascularization proceeds uneventfully. EBL 100ml. IVFs 1200ml. Surgical time = 3 hrs, 15 mins.

Will you extubate this patient following completion of the surgery?

If yes, will you extubate deep or wide-awake? Advantages? Disadvantages?

What are your specific concerns related to this emergence of this patient following revascularization?

In the PACU, the extubated child is alternating periods of agitation and combative behavior with periods of sedation and partial airway obstruction. Only a limited neurological assessment has been possible. P 90-100, BP 110-140/60-75, SaO₂ 92-98%.

What is your differential diagnosis of postoperative agitation in this child following this specific procedure?

How will you decide when this patient is ready for transfer to the PICU? If he is hemodynamically stable and sedated, should further neurological assessment be performed prior to transfer?

References

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