Introduction:
This case discusses the anesthetic management of moyamoya complicated by coarctation of the aorta and pulmonary hypertension in a 9 yr old.

Case Report:
A 9 yo M, ASA PS III, 28.2 kg with moyamoya disease (MMD) s/p cerebral bypasses s/t CVA referred for chest CT, brain MRI and cardiac catheterization after unsuccessful balloon angioplasty of coexisting coarctation of the aorta (CoA).
- Echo: normal biventricular function, mild residual CoA, moderate RVH with flattened IVS and tiny PFO shunting left to right
- Baseline NIBP was 124/65 and Hgb was 14.3
- Medications: aspirin and atenolol
- PE was unremarkable

Anesthetic management:
- Premedication: IV midazolam
- Induction: propofol, fentanyl and cis-atracurium
- Maintenance: sevoflurane ~1.5ET%, cis-atracurium, fentanyl
- ETCO₂ maintained mid 30’s
- Deep extubation w/ midazolam and dexmedetomidine

Cath results (Fig 2):
- Moderate pulmonary hypertension (PH), mPAP 50-60mmHg
- Rp 12 WUxM² w/ minimal response to NO
- LVEDP 13-15
- CoA gradient 7-10mmHg.

Chest CT w/ PH sequela and mid aortic syndrome. Stable brain MRI. Sildenafil and bosentan were started postoperatively and he was discharged home on POD 1 without sequelae. He has since undergone balloon dilation of PFO and medication adjustments.

Intraoperative Complications:
- Mild hypotension 80/40 during MRI responded to
  - 350 ml NS & 250 ml 5% albumin

Discussion:
MMD is a chronic cerebrovascular occlusive disorder usually affecting the Circle of Willis and resulting in collateral vascular network that resembles a “puff of smoke” (Fig.2), describing the cerebral angiographic findings.

Reports of extracranial lesions suggest a diffuse arteriopathy, as evidenced by its association with PH and renal arteriopathy. One report suggests a fibromuscular dysplasia etiology. MMD with CoA was reported in a patient with Noonan syndrome and MMD has been linked with other genetic abnormalities as well (i.e. neurofibromatosis, sickle cell anemia).

PH related to restrictive LV physiology is a predictor of early mortality in young adults with repaired CoA; however, no reports have shown MMD, CoA and PH to date. Anesthetic management needs to balance hemodynamic preservation while avoiding PH crisis and potential cerebral ischemia. Normothermia, normovolemia, normocapnia and normothermia are ideal. It is important to establish this balance prior to obtaining cardiac catheterization data.

Conclusion:
This is an unusual case of MMD with mild CoA and significant PH in a child demonstrating the challenges of anesthetic management.

References: