

## **Anesthetic Management of encephaloduroarteriosynangiosis (EDAS) for cerebral steno-occlusive angiopathy (Moya Moya Disease)**

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**Introduction:** Moya-Moya disease (MMD) is an ischemic cerebral vasculopathy with progressive stenosis of bilateral supraclinoid internal carotid arteries. Spontaneous formation of tortuous arterial collaterals re-establish cerebral blood flow from the basilar to the distal region, creating the characteristic findings of “puff of smoke” on a cerebral angiogram. Its occurrence may be familial (10 %) or sporadic. MMD is often a manifestation of a multisystem disease. It has a female preponderance (60%), and has an increased incidence in the Asian population. EDAS aims to re-establish blood flow to compromised areas of the brain. We present the anesthetic management of a patient undergoing bilateral EDAS procedure

**Case presentation:** A 15-year old female developed atypical Hemolytic Uremic Syndrome (HUS). A renal biopsy confirmed thrombotic renal angiopathy, requiring bilateral native nephrectomy to treat her severe hypertension. Four months following her nephrectomies, she presented with a dense left (L) hemiparesis and expressive aphasia. Diagnostic work up was significant for antecedent multiple ischemic vascular accidents. Magnetic resonance imaging (MRI) revealed subarachnoid infarctions in the right (R) centrum semi-ovale with possible subarachnoid hemorrhage (SAH) in the R frontal lobe and basal ganglia, as well as lack of flow in the R proximal anterior (ACA) and middle cerebral artery (MCA). Infarctions were also present in the L corona radiata, L caudate, and L external capsule. Cerebral angiogram confirmed stenoses of the L internal carotid (ICA), L MCA, as well as extremely tight stenosis of R ICA and MCA. Blood flow was compromised in both anterior cerebral arteries. The patient was on abdominal peritoneal dialysis (CAPD). Her medications included: iron, calcium gluconate, prednisone, sodium bicarbonate, clonidine, nicardipine, lovenox, ranitidine, hydralazine, and epogen. She underwent a two-staged revascularization procedure. Her first EDAS addressed disease in the right hemisphere three weeks after the onset of her left hemiparesis. In addition to standard ASA monitors, an indwelling arterial catheter as well as somatosensory evoked potentials (SSEP) and continuous electroencephalography (EEG) were employed. A CVP was deferred due to her bleeding diathesis. Her uremic bleeding diathesis was managed with DDAVP. General anesthesia was induced using etomidate, cisatracurium, and fentanyl. Maintenance consisted of remifentanyl and propofol infusions. Her Intra-arterial blood pressures were maintained between 130/70 at all times. No new fixed deficits were noted at the conclusion of her right EDAS. After a three-week interval a left EDAS was performed. During that time her anti-hypertensive medications had been adjusted. On the day she underwent CAPD in preparation for her second operation. Following her CAPD, she was noted to be hypotensive (unobtainable BP) and unresponsive. She responded to the administration of 1500 ml of normal saline and 250 ml 5% albumin with her blood pressure returning to 110/60. In an attempt to avert a fixed deficit, it was decided to proceed with surgery. Despite careful attention to BP management intra-operatively, decreased R median L cortical SSEP's were noted. In contrast to her first procedure anesthetic requirements were modest. Following her procedure a new right hemiparesis and mutism were present. Cranial tomography (CT) revealed a left cortical CVA.

**Discussion:** The surgical treatment of MMD is much debated. Since its original description in 1957, EDAS and superficial temporal artery-middle cerebral artery anastomosis with encephalo-duro-arterio-myo-synangiosis (STA-MCA EDAMS) have been the available surgical options. The latter carries great promise because blood flow to both the MCA and ACA territories is potentially restored. Careful attention to intravascular volume is a main factor in the success of this procedure. Fluid repletion in patients on dialysis is challenging. In our case, transesophageal echocardiography, a reliable indicator in a stiff myocardium of intracavitary filling might have been helpful to guide fluid management. Thereby excluding fluid depletion as a reason for her new neurological deficits.

### **References:**

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