MCC-7173

Anesthetic Management of a Pediatric Patient with Acute Stroke and New Onset Restrictive Cardiomyopathy

Primary Author: John-Paul Sara MD
Westchester Medical Center

Co-Authors: Irim Salik, MD;

Introduction

Stroke in the pediatric population is a rare occurrence, tends to be poorly understood, and has varying etiologies. Diagnosis requires careful clinical examination in conjunction with brain imaging. Most children experience neurologic deficits with long-term treatments focused on physical, developmental, and psychosocial complications. We present the case of a pediatric patient who presented with an acute stroke and was subsequently found to have an underlying restrictive cardiomyopathy.

Case & Anesthetic Course

A two-year-old, previously healthy and developmentally appropriate female presented to the Emergency Department with sudden onset right facial and right sided extremity weakness, drooling, and one episode of vomiting. Imaging with a Computed Tomography (CT) scan, a CT angiography, and a rapid MRI (Magnetic Resonance Imaging) showed indeterminate results clouded by motion artifact. Rheumatologic and hypercoagulability workups were negative.

A full Brain MRI and Magnetic Resonance Angiography (MRA) of the head and neck under anesthesia was requested. The patient, 14 kg, arrived to the MRI suite and standard ASA monitors were applied. The patient underwent an inhalational induction with Sevoflurane, nitrous oxide, and oxygen. Intravenous access was secured and the patient received a bolus of Propofol 50mg. The patient was intubated uneventfully with a 4.5 cuffed endotracheal tube. Sevoflurane was used at 1 MAC for maintenance of anesthesia during an approximately 2.5 hour anesthetic course, and dexmedetomidine 7 mcg was administered prior to emergence. During this anesthetic course, a complete transesophageal echocardiogram (TEE) was performed under anesthesia to properly visualize the left atrial appendage. The patient remained hemodynamically stable throughout the anesthetic course.

MRI/MRA showed an acute left middle cerebral artery infarct (slices to left side of attached image). TEE revealed severe bi-atrial enlargement with biventricular hypertrophy consistent with restrictive cardiomyopathy, as well as a patent foramen ovale. On the fourth day of admission the child’s neurologic status worsened with increased lethargy, confusion, and new onset seizures. A repeat MRI at this time revealed an acute right middle cerebral artery infarct. (slices on right side of attached image). On a subsequent TEE, the left atrial appendage demonstrated an echogenic mass. The neurologist surmised that the bilateral strokes were cardioembolic in nature.

Discussion
The incidence of pediatric acute ischemic stroke (AIS) is estimated at 2-13 per 100,000 per year in the first 5 years of life.1,2 Mortality is 2-11% but morbidity is 68-74%2 with known sequelae such as deficits in motor function, language, learning ability, behavior, and social skills presenting lifelong difficulty. The most common presenting symptoms are seizure (20-48%) or hemiparesis.2 Most episodes are idiopathic, but known associated conditions include congenital heart disease, infection, prothrombotic disorders, vasculopathies, and metabolic derangements.1-3 One prospective study identified cardiac disease as a risk factor for (AIS) in almost 1/3 of patients, and congenital structural defects were more common than acquired defects.3

Endovascular treatment of AIS remains the best option if feasible, however anesthetic management for this procedure remains controversial. General anesthesia may lead to worsened outcomes compared to sedation,4-6 although results are mixed. Anesthetic management of a patient with AIS remains focused on hemodynamic stability and maintenance of cerebral perfusion pressure (CPP), the primary determinant of cerebral blood flow. CPP is defined as the difference between mean arterial pressure and intracranial pressure. In the setting of ischemic stroke, thrombolysis may restore perfusion and decrease infarct size, but it may also lead to expansion of the infarct, edema, and hemorrhage. Hyperglycemia has been associated with worsened outcomes in a number of settings including stroke, TBI, acute coronary syndrome, and critical illness. Although there is a lack of evidence to support hypothermia in humans for cerebral protection, there is evidence that hyperthermia is associated with worsened outcome in settings of ischemic stroke, subarachnoid hemorrhage, cardiac arrest, and traumatic brain injury. In the setting of an acute stroke, hypotension and elevations in intracranial pressure should be avoided, the latter of which can occur during intubation without an appropriate depth of anesthesia.7

The incidence of Cardiomyopathy (CM) in children under 10 years old is 1.3 in 100,000. Restrictive CM (RCM) represents 2.5% of those (3.25 in 10,000,000). Prognosis is grim and two year survival from diagnosis is <50%.8 In this condition, impaired ventricular filling and increased left ventricular end diastolic pressure from a fibrotic myocardium cause a progressive increase in pulmonary vascular resistance (PVR) with concomitant decrease in stroke volume and cardiac output. Patients typically present late in the course due to compensatory adaptation until exercise tolerance severely decreases and are often found to have markedly elevated PVR. Massively dilated atria can lead to thromboembolic events. Idiopathic RCM is the most common etiology in children but known causes include amyloidosis, hemosiderosis, hypereosinophilia (Loffler's disease), endocardial fibroelastosis, and previous radiation.

During an anesthetic course of a patient with RCM it should be noted that bradycardia and hypovolemia should be avoided, as heart rate and preload remain the predominant determinants of cardiac output. Inotropic agents such as milrinone, dobutamine, and amrinone have been used successfully. Equally important is avoiding high airway pressures, hypercarbia, or hypoxia as all of these may cause an iatrogenic increase in PVR which may cause rapid hemodynamic decompensation when combined with decreased cardiac output.9

Despite these considerations anesthesia is considered safe for patients with cardiomyopathy. One retrospective study identified 4 cardiac arrests in 2 patients over 236 anesthetic events (1.7%) for children with cardiomyopathy during a 10 year period.9

Our patient presented with signs and symptoms of acute stroke, a rarity in the pediatric population. During the initial anesthetic course it was discovered that this was a sentinel complication of
idiopathic restrictive cardiomyopathy. Reassuringly she did not present with hemodynamic instability or bradycardia, so our intent of maintaining hemodynamic stability and cerebral perfusion pressure throughout the anesthetic course was not compromised. Over the course of several subsequent anesthetics, our team took her conditions into consideration and adjusted goal parameters for blood pressure, heart rate, and volume balance accordingly to optimize the patient's clinical outcome.


