Anesthetic Management of a Pregnant Woman with Sickle Cell Disease and Coexisting Moyamoya Disease: a case report

Presenting Author: Allen E Coleman MD
Presenting Author's Institution: Wayne State/Detroit Medical Center - Detroit, Michigan
Co-Authors: Tatayana Tenenboym CRNA - Wayne State/Detroit Medical Center - Detroit, Michigan
Vitaly Soskin MD, PhD - Wayne State/Detroit Medical Center - Detroit, Michigan

Introduction: Anesthetic management of a parturient with coexisting Sickle Cell (SCD) and Moyamoya disease (MMD) aims to maintain normal physiologic parameters with regards to blood pressure (BP), cerebral blood flow (CBF), temperature, carbon dioxide, and oxygen saturation to avoid the potential development of severe disease specific complications.

Case Report: A 20 years old African American patient at 35 weeks gestation with coexisting SCD and MMD was admitted with acute chest syndrome. Her condition acutely deteriorated with the development of hypoxia, anemia, and congestive heart failure, necessitating blood transfusion and urgent cesarean delivery. Surgical management included placement of an epidural catheter, arterial line, central line, and all other standard American Society of Anesthesia monitors. A viable male infant was delivered with APGAR scores of 8 and 9 at 1 and 5 minutes, respectively. Following delivery the patient's condition significantly improved without the occurrence of any postoperative complications. This allowed discontinuation of the epidural catheter and invasive monitors after 24 hours.

Discussion: Maintenance of BP, normothermia and arterial carbon dioxide within strict parameters is critical in these patient populations. Avoidance of hyperventilation is critical in MMD as reduced CBF may precipitate cerebral ischemia while hypoventilation in SCD, leading to hypercapnia and acidosis may induce sickle cell crisis. As nearly all induction agents used in general anesthesia typically produce a dose dependent decrease in CBF and volatile anesthetics are cerebral vasodilators, their use may result in an occurrence of cerebral steal in MMD or SCD. Normal body temperature must be maintained as hypothermia can lead to cerebral vasospasm in MMD and vasoocclusive crisis in SCD. In this case we have shown that epidural anesthesia along with standard and invasive monitors can allow carefully maintained physiologic control and provide for excellent operative and postoperative pain control in patients with both SCD and MMD.