A Tale of Two Hemoglobins: Management of a Parturient with Suspected Congenital Methemoglobinemia

Abstract Type: Case Report/Case Series
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Introduction: Methemoglobinemia is a congenital or acquired (e.g., from medications) disorder resulting from oxidation of Fe2+ to Fe3+ (which is unable to bind oxygen). Affinity of Fe2+ for oxygen is increased, impairing oxygen release. Surgery or labor may provoke congenital forms, where methemoglobin reductase is reduced. Management in a parturient with congenital methemoglobinemia presents additional challenges, since either surgery or labor are required for birth, and in a methemoglobin (MetHb) crisis, fetal non-wellbeing may be an early sign, needing rapid delivery.

Case Report: A 25-year-old G1P0 female was seen prior to delivery regarding a history of suspected congenital methemoglobinemia. Four year prior, she was diagnosed after a single cyanotic/hypoxic episode following a biopsy. Lidocaine was used, and she desaturated to SpO2 of 82% with PaO2 of 84 mmHg. A MetHb level of 13.3% was found, and follow-up testing showed a low level of MetHb-reductase (6.3 IU, normal 8.2-19.2 IU), normal MetHb (0.5%), and a normal lactate (1.5 mmol/L). Repeat MetHb was 8.4 IU. It was deduced that with her enzyme at the lower end of normal, she could be more susceptible to oxidative stress, and a conservative plan was formed for vaginal delivery under neuraxial analgesia with continuous fetal monitoring during labor, and MetHb levels every two hours. Fetal distress would trigger transfusion of packed red blood cells (free of MetHb) to increase oxygen delivery, methylene blue to reduce MetHb present, and then to proceed with urgent Cesarean delivery should these prove unsuccessful. Chloroprocaine and bupivacaine were tentatively approved for use due to lack of literature, with caution against using lidocaine.

During her labor, an epidural was placed using chloroprocaine for local infiltration, and was test-dosed with 15 mcg of epinephrine and 2.5 mg of bupivacaine. Initial MetHb levels were 0.2% (normal: 0.4-1.5%). Her MetHb levels stayed normal during labor (peak of 1.5% during pushing efforts). Fetal status was reassuring throughout, but delivery was complicated by shoulder dystocia and second degree laceration. She inadvertently received an epidural lidocaine bolus for repair, but a MetHb level checked immediately after this had fallen back to 0.2%. She recovered uneventfully.

Discussion: Management of a parturient with congenital methemoglobinemia presents challenges, due to unavoidable triggering of MetHb formation (i.e., labor or surgery), leading to fetal hypoxia. Classical teaching dictates avoidance of prilocaine and benzocaine, and there are few cases of lidocaine was a triggering agent. As neuraxial anesthesia is the mainstay of OB anesthesia, and as it is unclear whether esters or amides are riskier in patients, with a paucity of literature, certainly more research could be done to evaluate triggering potential of these common anesthetic agents.

Reference: