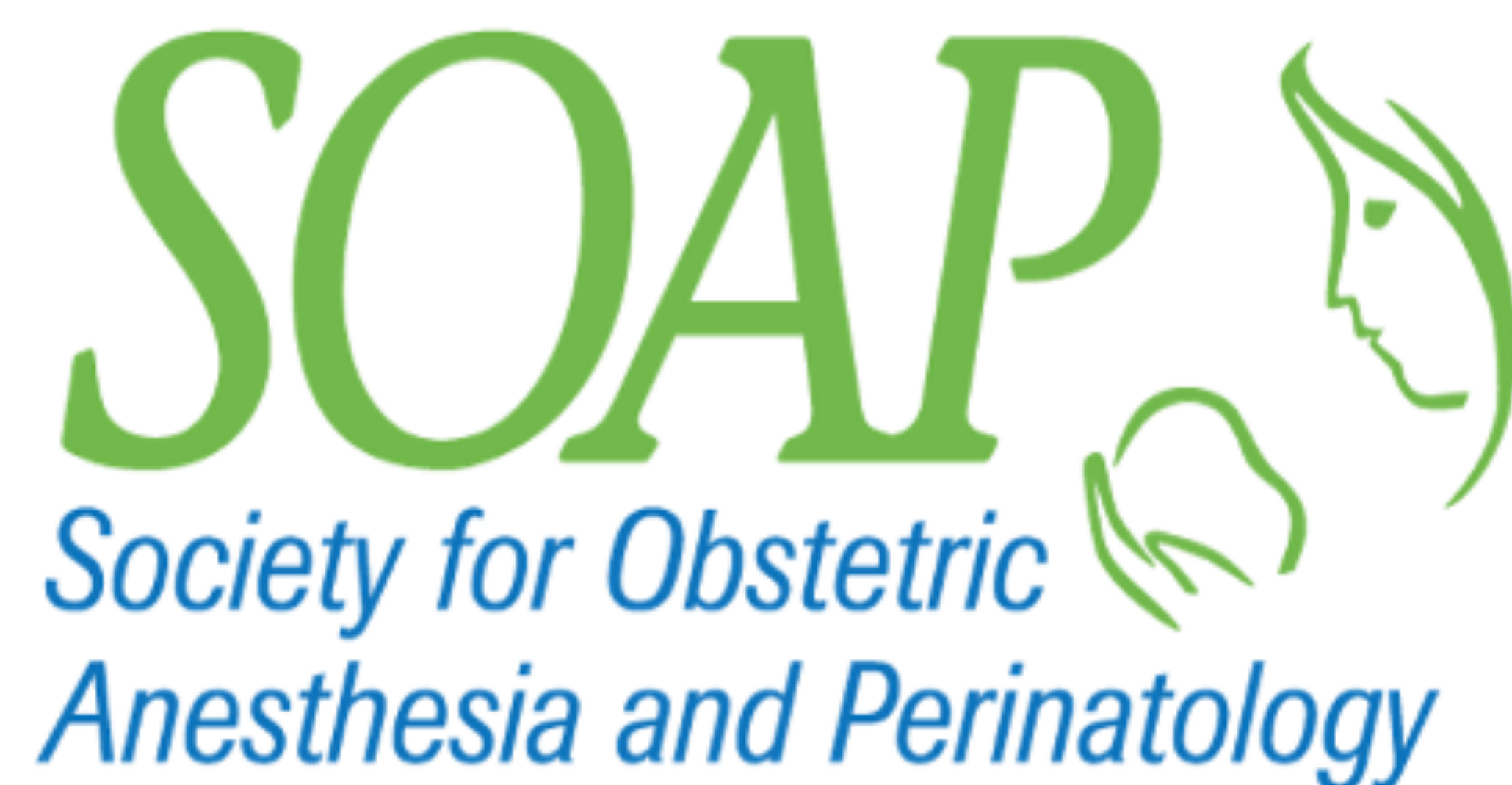


Sickle Cell Crisis and Aortic Stenosis

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Introduction

- Sickle cell disease (SCD) is an autosomal recessive disorder that affects approximately 100,000 people in the US.
- Manifestations of SCD are exacerbated by pregnancy causing a higher frequency of vaso-occlusive crises, worsening anemia and hypercoagulability.
- These hematologic changes can increase metabolic demand and cardiac work exacerbating pre-existing cardiac valve abnormalities, most notably aortic stenosis.

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Case Presentation

- 23 year old G1P0 at 31w4d with a mhx of sickle cell disease, asthma, pulmonic stenosis s/p valvuloplasty as an infant, moderate aortic stenosis and SVC syndrome presents with complaints of chest pain, lower extremity pain, shortness of breath and neck swelling. Spo2 was 85% which improved with supplemental O2. Pt admitted for pain control.
- TTE: normal systolic and diastolic function with moderate to severe aortic valve stenosis.
- At 34w4d pt had numerous episodes of sinus tachycardia associated with worsening shortness of breath and chest pain. Workup showed decreased hematocrit from baseline. The patient was placed on supplemental O2 and the decision was made to proceed with delivery at 35w via C-section.
- One unit of pRBC administered. Epidural placed and a right radial arterial line. 20mL of 2% Lidocaine was administered.
- Supplemental oxygen was applied via nasal cannula and a bair hugger was applied.
- After delivery of the infant, there was an increase in the heart rate up to 115 which improved after administration of esmolol, all other vital signs remained normal.

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Discussion

- During a sickle cell crisis, treatment is targeted to decrease the rate of sickling in the vasculature.
- Hypertension and tachycardia often occur during these pain crises and are poorly tolerated in stenotic cardiac lesions.
- Non pharmacologic steps include aggressive hydration, supplemental oxygen and maintaining normothermia.
 - In a patient with preexisting moderate to severe AS, this can pose a challenge as excess hydration can exacerbate the valvular pathology in conjunction with the increase in blood volume and cardiac output during pregnancy.
- During delivery for patients with both pathologies, it is imperative to optimize pain control, volume status and monitor hemodynamic status closely to quickly treat any sudden variations.