



Delivery Planning in a Parturient with End Stage Systemic Sclerosis: When a General Anesthesia is Not an Option

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Introduction

- Systemic sclerosis (SS) is a chronic autoimmune connective tissue disorder characterized by cutaneous and multiple organ-base fibrosis resulting in progressive end-organ dysfunction.
- It has an incidence of 2–10 cases per million and is three to five times more common in women than men.
- SS may occur in women of childbearing age, and pregnancies in this population are associated with a increased risk of adverse obstetric and maternal outcomes.
- SS results in significant challenges for the practicing anesthesiologist due to its rarity, multi-system involvement, and limited evidence-based guidance for optimal perioperative care

Case Presentation

- 37 year old G7P1 at 34+4weeks with severe SS and failure to thrive admitted for induction of labor and care coordination.
- Manifestations of her SS included: paroxysmal supraventricular tachycardia (SVT), mitral valve prolapse, tricuspid regurgitation, apical aneurysmal dilation of her heart, recurrent pericardial effusions (including a history of tamponade requiring pericardiocentesis), interstitial lung disease, eosinophilic esophagitis, and significant cutaneous involvement including difficult IV access and contractures.
- Anesthetic history was notable for difficult IV access.
- Her pregnancy was notable for worsening SVTs, including a cardiac care unit admission at 30+4weeks complicated by pre-term labor.

- Based on her exam (Figure 1), it was uncertain if intubation or even mask ventilation would be possible. It was also unclear if she had enough laxity in her vaginal tissue to allow for a vaginal delivery, yet, if she went for a c-section, surgical closure would be difficult due to the limited laxity.
- Multiple meetings were held between Maternal-fetal-medicine (MFM), nursing and obstetric anesthesiology. Given her history of difficult IV access, an US-guided triple lumen catheter was placed prior to her labor induction. An US assisted epidural was performed prior to starting any labor medications. Positioning for neuraxial was challenging, but an L3-4 epidural was uneventfully placed. Advanced airway equipment (including pediatric indirect laryngoscopes of multiple sizes, medications for airway topicalization, and a flexible bronchoscope) was available on the labor floor, and plans were made to call a hospital difficult airway code if she needed a general anesthetic. She progressed quickly once her induction was started. She was brought to the operating room to push, and a vacuum was used to assist the vaginal delivery of a 1750gm baby.



Figure 1. Sclerotic changes: Microstomia and mask-like facial stiffness.

Discussion

- Pregnancy in SS may be uneventful, with both good maternal and fetal outcomes. However, careful antenatal evaluations, discussion of potential problems, and participation in a high-risk obstetric monitoring program is very important to optimize the best outcomes.
- Skin contractures and microstomia make tracheal intubation difficult, while esophageal dysmotility and sphincter incompetence can lead to aspiration.
- Neuraxial anesthesia is safe in patients with SS, particularly with ultrasound guidance and allows the most benefit during delivery. It not only provides adequate anesthesia but also provides peripheral vasodilatation and increased skin perfusion of lower extremities.
- This case illustrated the importance of multidisciplinary communication and risk identification and mitigation in patients with severe SS. Open communication regarding obstetric and anesthesiologic concerns were key to developing a safe delivery plan along with appropriate contingencies.

References

1. Carr, Z.J., Klick, J., McDowell, B.J. et al. An Update on Systemic Sclerosis and its Perioperative Management. *Curr Anesthesiol Rep* 10, 512–521 (2020). <https://doi.org/10.1007/s40140-020-00411-8>.
2. Steen VD. Scleroderma and pregnancy. *Rheum Dis Clin North Am*. 1997 Feb;23(1):133-47. doi: 10.1016/s0889-857x(05)70319-7. PMID: 9031379.
3. Clark KE, Etomi O, Ong VH. Systemic sclerosis in pregnancy. *Obstet Med*. 2020;13(3):105-111. doi:10.1177/1753495X19878042.
4. Yang L, Sun W, Yao Q. Management of pregnancy with diffuse cutaneous systemic sclerosis: a case report and literature review. *J Int Med Res*. 2021;49(11):3000605211060160. doi:10.1177/03000605211060160