

Intentional Minimalism: Anesthetic Choice for Cesarean Section in Patients with Myotonic Dystrophy

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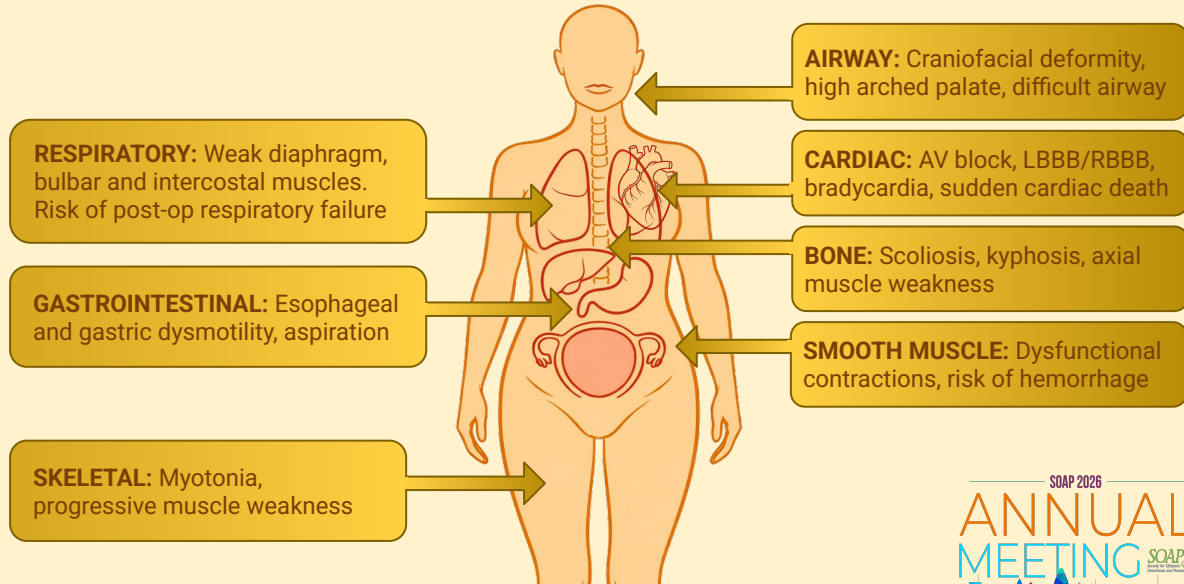
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Introduction

BACKGROUND

- Myotonic dystrophy 1 (DM1) is a multisystem genetic disorder affecting roughly 1 in 8,000 individuals worldwide¹
- Fertility is slightly reduced in this population¹
- Pregnant patients with DM1 have higher reported rates of miscarriage and preterm birth¹
- Given the multisystem nature of DM1, patients are considered high risk from an obstetric and anesthetic perspective

MYOTONIC DYSTROPHY



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Case Presentation

Patient: 34 year old G4P3003 with a history of three c-sections, PPH, prior cardiac arrest, myotonic dystrophy



Patient presents to OSH at 28w0d after mechanical fall, noted to be hypoxic



XR and chest CT revealed PNA and dilated esophagus (suspected GI dysmotility)



Airway secured for PEG/bronchoscopy via awake fiberoptic due to respiratory status and a MP IV exam



Remained intubated for one day after successful PEG placement. PNA treated with antibiotics, and discharged on room air



Presented to L&D at 37w0d for scheduled c-section. On exam, frail-appearing with short stature and airway exam was notable for limited mouth opening.



Delivered uneventfully via cesarean, with a **primary anesthetic* of low dose CSE, opioid-only spinal, incremental lidocaine dosing**

ANESTHETIC CHOICES*:

- Opioid-only spinal
 - 75 mcg intrathecal morphine, 15 mcg intrathecal fentanyl
- Low-dose combined spinal epidural
 - Incrementally dosed with 2% lidocaine (total 19 mL)
- Prophylactic methylergonovine and oxytocin were administered immediately after delivery of the newborn.
- Carboprost, magnesium, and succinylcholine were avoided
- Intraoperative discomfort treated with 3% chlorprocaine on the field, + 3 mL of 2% lidocaine via epidural + 0.5 mg IV midazolam.
- Low-dose PCEA for postoperative pain control
- Received a second dose of neuraxial morphine before epidural removal on POD 1

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Discussion & Key Points

- The multisystem nature of DM1 requires vigilant anesthetic planning
- Despite attention to hemodynamics and respiratory status, the patient still developed pulmonary edema, which resolved on POD2
- The primary goals in this case were:
 - Avoid weakening the intercostal and accessory muscles with a full-dose spinal
 - Avoid a general anesthetic given her stature, poor airway exam, respiratory complications, and vulnerability to certain IV paralytics and sedatives.
- Rescue airway equipment included a glidescope, McGrath 2 blade, 5.0-7.0 ETT sizes, and pediatric oral airways.
- Other considerations were aspiration risk, hemorrhage, and myotonic crisis, which can be triggered both pregnancy and by perioperative stimuli (pain, shivering, cautery)^{2,3}.
- Anesthetic planning for pregnant patients with DM1 should reflect intentional minimalism: emphasizing avoidance of triggers rather than escalation of therapy.



Figure 1: Postoperative CXR on POD1 with patchy atelectasis in right lower lung and retrocardiac

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2. Paterson, R. A., Tousignant, M., & Skene, D. S. (1985). Caesarean section for twins in a patient with myotonic dystrophy. *Canadian Anaesthetists' Society journal*, 32(4), 418–421.

3. Owen, P. M., & Chu, C. (2011). Emergency caesarean section in a patient with myotonic dystrophy: a case of failed postoperative extubation in a patient with mild disease. *Anaesthesia and intensive care*, 39(2), 293–298.