

# Anesthetic Considerations in Spinal Muscular Atrophy Type II for Cesarean Delivery



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#### **BACKGROUND**

#### Spinal muscular atrophy (SMA)

- Defect in the survival motor neuron-1 (SMN-1) gene
- Variable phenotypes
- Improvements in treatment → more women reaching child-bearing age

#### **Pregnancy and SMA**

- Increased risks of pre-term labor
- Worsening of baseline respiratory weakness
- Scoliosis & difficult with neuraxial placement<sup>2</sup>

# **SMA Subtypes**

- I. Infantile onset, respiratory distress, inability to sit upright
- II. 6-10mo onset, wheelchair-bound
- III. 18m-adulthood onset, independently mobile
- IV. Late-onset after age 30, maintain their mobility<sup>1</sup>



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#### **CASE REPORT**

#### Presentation

- 35 year-old female G3P1102 at 37w0d with SMA type II
- Wheelchair-dependence
- Scoliosis s/p thoracolumbar spinal fusion
- Restrictive lung disease/BiPAP
- Obstetric hx: 2 prior cesarian deliveries under GA with LMA
- Physical exam: Mallampati
  4, reduced oral opening
  & thyromental distance,
  reduced cervical motion,
  severe limb contractures



- Plan: Cesarean delivery general TIVA with ETT
- <u>Induction</u>: propofol 200mg, fentanyl 25mcg, lidocaine 50mg, propofol infusion
- <u>Intubation</u>: RSI with video laryngoscope grade IIb view 6.5 ETT
- Ventilation: Controlled

   → PSV → manual with

   TV ~245mL → extubation
- Pain management: TAP blocks, hydromorphone 1.0mg IV, ketorolac 15mg IV

#### Postop

- <u>POD 0</u>:
  - Immediate hemorrhage
  - 300cc clot in vaginal canal (atony)
  - Rectal misoprostol 800mcg, methergine 0.2mg IM
- <u>POD 1</u>:
  - H/H  $11.2/35 \rightarrow 6.6/21$
  - 1U pRBC and iron
  - Persistent bleed
    → methergine & TXA
  - Pain control: MMPR insufficient → PCA pump
- <u>POD 3</u>:
- Discharged home





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### **LEARNING POINTS**

Risks of Vaginal Delivery

- Abdominal muscle weakness (limits pushing)
- Prolonged immobility → reduction in pelvic diameter
- Neuraxial feasibility

Risks of Cesarean Delivery (Spinal)

- Reductions in FEV1 and VC after spinal placement
- Scoliosis (difficult/impossible placement)

Risks of Cesarean Delivery (General)

- Aspiration risk, difficult airway
- Respiratory muscular weakness and increased difficulty to wean from mechanical ventilation
- Avoid paralysis
- ETT vs. LMA: weigh aspiration risk

#### References

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