

Interdisciplinary Management of a Parturient with Type II Spinal Muscular Atrophy

Mikayla Troughton, MD; Feyce Peralta MD; Mahesh Vaidyanathan MD, MBA; Samantha Lu, MD
Department of Anesthesiology, Northwestern University Feinberg School of Medicine, Chicago, IL

Background

- Spinal Muscular Atrophy (SMA) is an autosomal recessive disorder of the SMN1 gene, characterized by muscle weakness and atrophy¹
- Disease classification: SMA Type I (most severe) – SMA Type IV (least severe)
- Oral risdiplam and intrathecal nusinersen are allowing women with severe forms of SMA to survive into their reproductive years
- In 2018, a total of 18 cases of pregnancy in those with SMA Type II had been described²
- We report a successful primary cesarean delivery in a patient with SMA Type II and severe restrictive lung disease

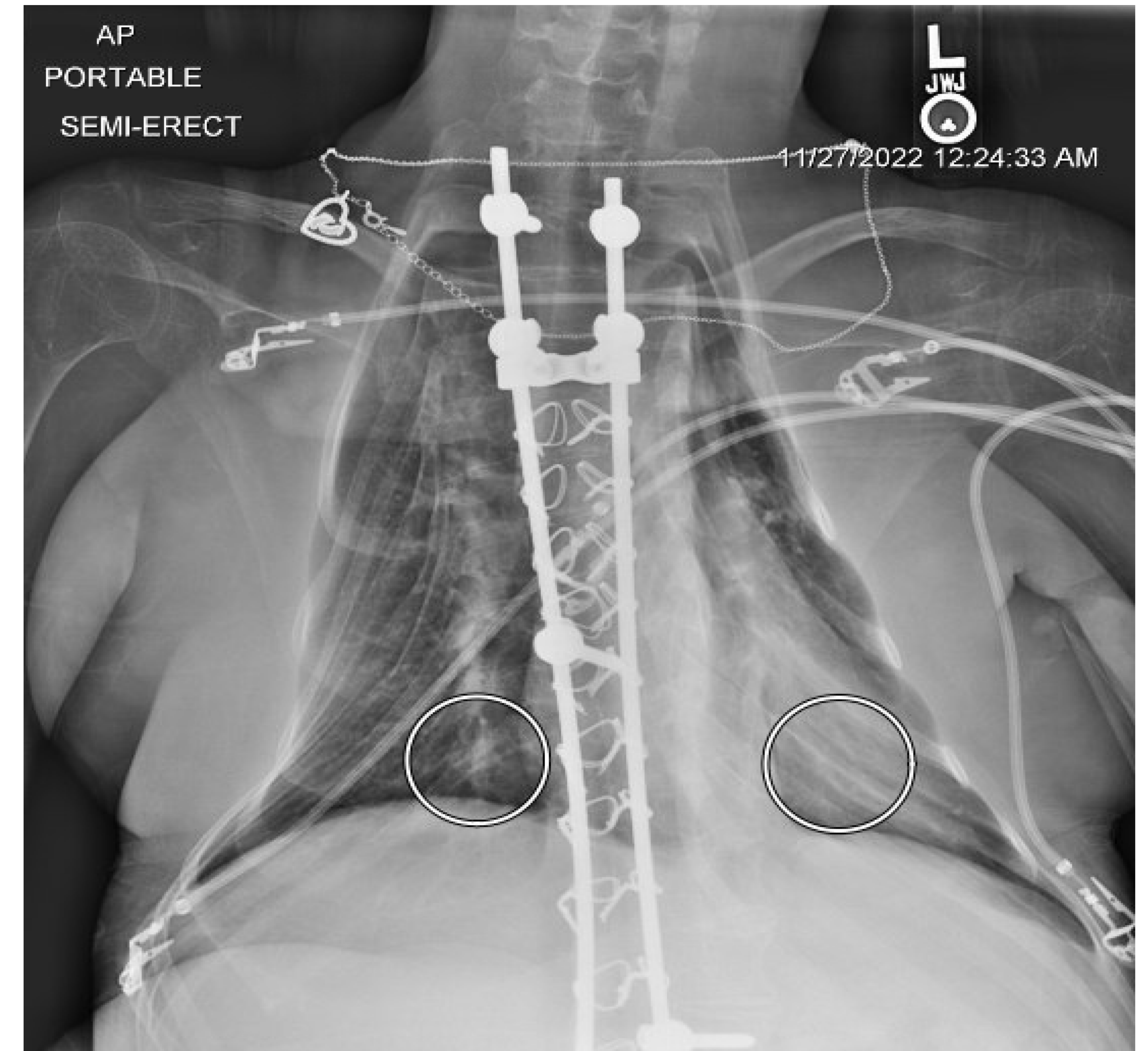
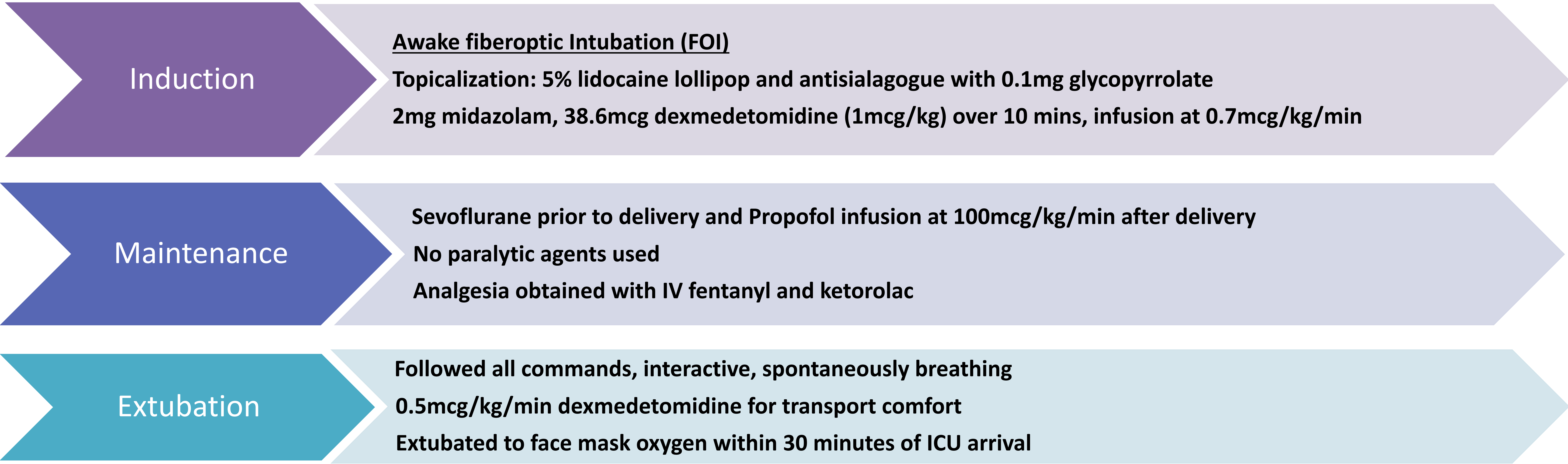


Figure 1. Second trimester chest XR of our patient

Case Presentation

- 29 yo G1P0, PMHx type II SMA, PSHx Harrington rod placement, scheduled for primary cesarean delivery at 34w2d
- Studies: PFTs - severe restrictive lung disease (FEV1 25%, FVC 23% of predicted), BiPAP up to 12 hours/day
- Physical Exam: Wheelchair bound, bulbar involvement, dysphagia, severe hip contractures, 0/5 motor for bilateral lower extremities
- Airway: Mallampati II, limited mouth opening, TM distance <3FB

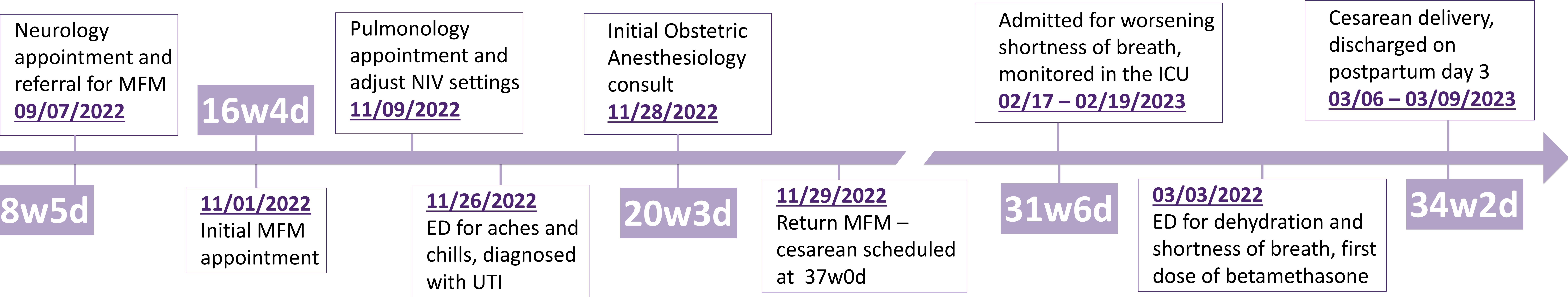


Discussion

- SMA patients are more likely to have respiratory compromise leading to preterm delivery²
- Given severe respiratory compromise, dysphagia, inability to manage secretions, difficulty with positioning, and history of Harrington Rods with residual scoliosis, we chose to perform **general anesthesia with an awake fiberoptic intubation**
- Depolarizing muscle relaxants are **contraindicated**, and SMA patients are sensitive to non-depolarizing agents
- Patient was discharged home on postpartum day 3, baby was discharged from the NICU on day 25 of life

Recommendations

- Multidisciplinary care coordination



References:
1) Nunes Vicente (2021) Neurological Sciences 42:9
2) Abati (2018) Journal of the Neurological Sciences 388:50-60