

Pediatric Patient with Spinal Muscular Atrophy Type 1 Undergoes Tracheostomy and PEG Tube Placement

Angela Suh, DO; Alex Mohtadi, DO; Evan Salant, MD

BACKGROUND

- Spinal Muscular Atrophy (SMA) is an autosomal recessive neurodegenerative disorder characterized by progressive loss of alpha motor neurons in the spinal cord, leading to muscle weakness and trouble breathing, coughing, and swallowing. Type 1 is most severe, presenting before 6 months of age [1].
- Patients present with recurrent respiratory infections, prolonged oxygen requirement, and BiPAP/NIPPV dependence. Airway hypotonia and bulbar dysfunction increase risk of upper airway collapse and aspiration [2].
- Succinylcholine is contraindicated due to risk of life-threatening hyperkalemia from motor neuron denervation. Non-depolarizing agents (rocuronium) are preferred to avoid exaggerated NMBA response in this population [3].

HPI

- 2 yo male with SMA Type 1, poor weight gain, global developmental delay was admitted for acute hypercarbic respiratory failure secondary to aspiration pneumonia and atelectasis.
- Initial blood gas showed severe respiratory acidosis (pH 7.14, CO2 80). Patient had been previously cleared for oral feeding following outpatient swallow study, but chest x-rays showed worsening of LLL consolidation after resuming PO intake.
- Patient admitted to the PICU for non-invasive positive pressure ventilation (NIPPV) and nasogastric tube feeds initiated to minimize aspiration
- Pediatric surgery was consulted and decision was made to undergo simultaneous percutaneous gastric tube and tracheostomy tube placement after failure to wean patient from NIPPV.

PHYSICAL EXAM

- On exam, patient was hypotonic, weak, cachectic, with pectus excavatum, scaphoid abdomen, and scoliosis.
- Weight at time of surgery: 6.2 kg
- He arrived to the OR with a 24 gauge IV in the L hand, NG tube in place.

INTRAOPERATIVE COURSE

- Patient preoxygenated with 100% O2 via face mask.
- Induced with 20 mg propofol, 6 mg rocuronium, 10 mcg of fentanyl for intubation.
- Laryngoscopy was performed with a Glidescope Miller S1, CMAC S0, and then multiple Fiberoptic attempts were made with successful intubation on the fifth attempt.
- Intubated with size 3.5 single-lumen cuffed endotracheal tube.
- NMBA prolongation was not a concern given planned tracheostomy placement and postoperative mechanical ventilation.

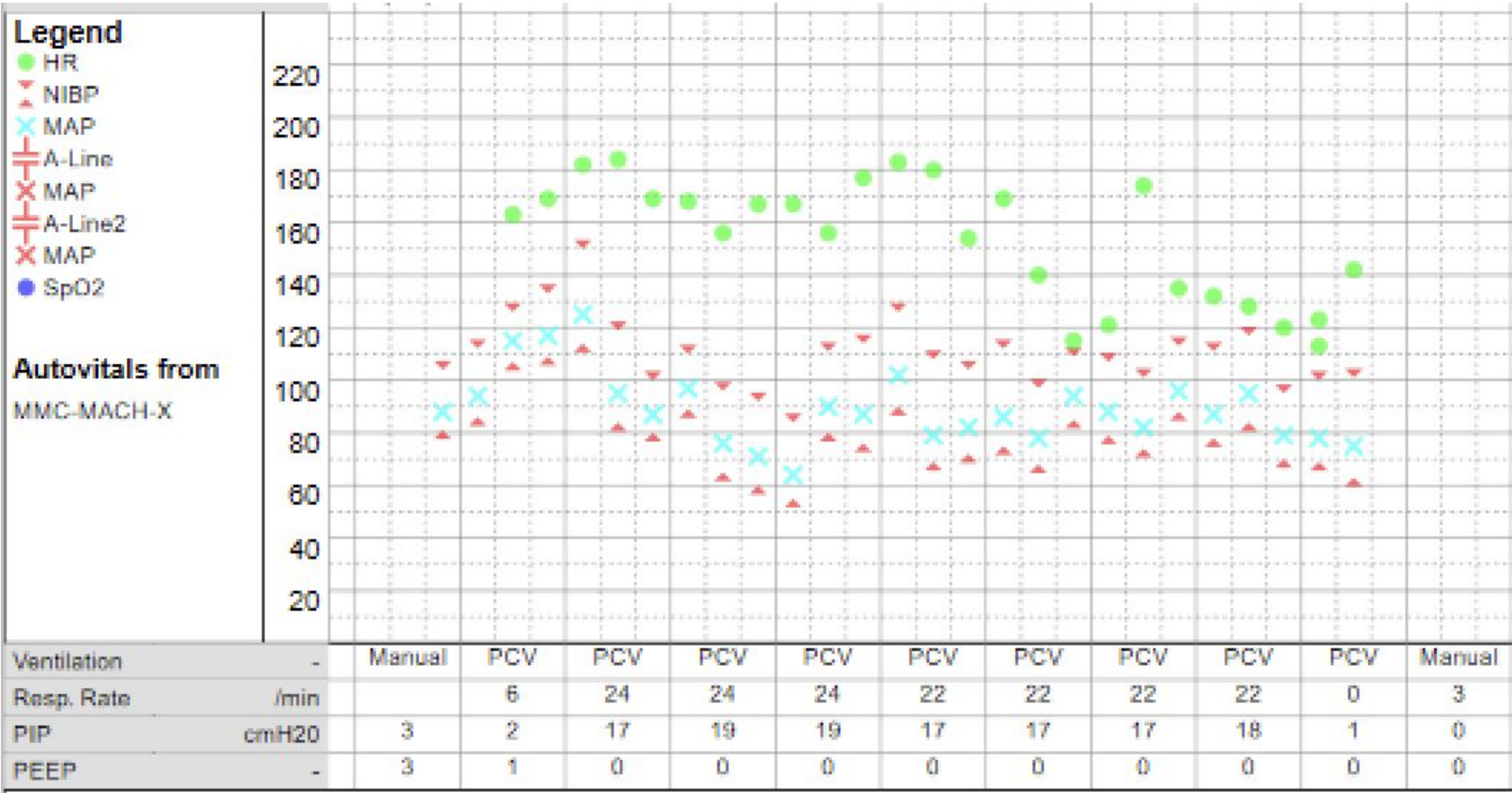


Figure 1: Vital signs throughout procedure

POSTOPERATIVE COURSE

- The patient underwent a successful procedure and had an uneventful transport to the PICU on monitor and ambu bag via tracheostomy tube.
- During transport, he was noted to have spontaneous breathing with no respiratory distress.
- In the PICU, he was eventually weaned from sedation and ventilator support and discharged from the hospital.



Figure 2: Postoperative chest x-ray showing new tracheostomy and PEG tube

CONCLUSION

- Patients with SMA are often considered to have compromised airways due to poor pharyngeal tone.
- Succinylcholine use is contraindicated due to risk of life-threatening hyperkalemia.

References:
[1] National Institute of Neurological Disorders and Stroke. "Spinal Muscular Atrophy." www.ninds.nih.gov, 23 Jan 2023.
[2] Kang, S-W. "Pulmonary Rehabilitation in Patients with Neuromuscular Disease." Yonsei Medical Journal, vol. 47, no. 3, 2006, p. 307.
[3] Martyn, JA et al. "Succinylcholine-Induced Hyperkalemia in Acquired Pathologic States." Anesthesiology, vol. 104, no. 1, 2006, pp. 158-169.