

Anesthetic Management of Postpartum Patient with Huntington’s Disease

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Introduction

- Huntington’s Disease (HD) is a neurodegenerative disease
- Autosomal dominant inheritance
- Affects 10-15 per 100,000 people of European descent
- Psychiatric symptoms typically manifest first, followed by random, uncontrollable jerky movements known as chorea
- Severe chorea can limit modes of anesthesia
- There is little literature describing the anesthetic considerations for peri-partum patients with HD
- We describe a case in which a patient with suspected HD was successfully managed for a bilateral tubal ligation (BTL)

Presentation

- 32yo G3P3 presented for open BTL after uncomplicated spontaneous vaginal delivery without anesthesia
- History of worsening choreiform movements over the past few years, likely HD, who is otherwise healthy
- Strong family history of chorea with her father who passed at 35yo and 6 of 9 siblings demonstrating symptoms as well
- She refused referrals for genetic counseling and testing
- Neurology notes reporting worsening symptoms
- On exam, she demonstrated dysarthria, flycatcher’s tongue, wide based, unsteady gait, and diffuse chorea in her head, trunk, and all extremities

Preoperative Considerations

- Although BTLs are commonly done under spinal at our institution, the patient’s involuntary movements were concerning for risk of injury
- A plan was made for GETA

Intraoperative Management

- Positioned on the operating bed with safety straps to limit her chorea and prevent fall
- She was pre-oxygenated and an RSI induction was performed
 - Fentanyl 100 mcg
 - Lidocaine 50 mg
 - Propofol 2 mg/kg
 - Rocuronium 1.2 mg/kg
- She was intubated with a 6.0 ETT using a Mac 3 blade
- Anesthesia was maintained with sevoflurane and supplemented with dexamethasone, hydromorphone, and ondansetron
- The procedure was uneventful, lasting 71 minutes
- Neuromuscular blockade was reversed with neostigmine (40 mcg/kg) and glycopyrrolate after 2 out of 4 twitches had returned at the adductor pollicis
- After 10 minutes, she had 4 twitches and sustained tetanus with adequate spontaneous tidal volumes, and she was extubated

Chorea vs. Residual Paralytic

- Shortly after extubation, the patient’s head and limbs exhibited uncoordinated and non-purposeful movement
- It was difficult to distinguish between baseline chorea versus a “floppy fish” appearance from residual neuromuscular blockade
- The patient seemed weak and unable to phonate
- An additional dose of neostigmine and glycopyrrolate was given with significant improvement
- She was transported to PACU with supplemental oxygen and an uneventful postoperative course.

Discussion

- There are several physiological and pharmacological considerations for HD patients undergoing GETA
- Case reports have shown an exaggerated response and prolonged apnea following succinylcholine
- As in our case, return of chorea on emergence may confound adequate neuromuscular blockade reversal
- Agents such as sugammadex may be considered more appropriate for HD patients where there is a question of adequate reversal
- Aspiration risk is increased in HD patient with dysphagia
- Medications that offset the balance of dopamine and acetylcholine in the striatum may worsen chorea
- Patients with severe HD are often on several psychiatric medications which may limit medications intraoperatively
- Location for testing train-of-four should be chosen carefully