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CLINICAL VIGNETTE

Anesthetic Considerations in a Patient with Type III Spinal Muscular Atrophy

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Background

Spinal Muscular Atrophy (SMA) is a monogenic neuromuscular junction disorder characterized by variable levels of progressive muscle weakness and atrophy in childhood and adolescence and is the most common monogenic cause of infant mortality in the world.[1] The disease is caused by autosomal recessive mutations in the survival motor neuron 1 (SMN1) gene located on chromosome 5q13.2, which results in gene deletion or mutations that interfere with mRNA synthesis in motor neurons that ultimately cause cell death of the anterior horn motor neurons of the spinal cord.[2]

Types I and II SMA are more severe presentations of the disease. Classically, type I presents in infancy while type II presents between 3 to 15 months of age. Both variants of the disease present with progressive, severe respiratory and global motor deficits. These patients develop significant respiratory weakness and may require mechanical ventilation. Furthermore, weakness of the bulbar muscles causes difficulty feeding, failure to thrive and pooling of secretions.[3]

Type III SMA is characterized by juvenile onset usually from ages 18 months to early adulthood, with progressive proximal lower extremity weakness and variable levels of upper extremity weakness, manifesting as frequent falls and difficulties with climbing stairs in early childhood.[3] Most patients eventually become wheelchair bound; however, SMA type III is not associated with scoliosis or respiratory muscle weakness, and they live a normal lifespan.[3]

Traditional therapy for SMA is supportive with a focus on nutritional supplementation, respiratory support, and physical therapy, which have been shown to be variably effective in preventing failure to thrive, aspiration, and the need for mechanical ventilation in patients with severe SMA[4]. Respiratory support primarily involves mobilization and clearance of airway secretions while chest physiotherapy and non-invasive ventilation improve quality of life for patients with respiratory failure.[4]

For the anesthesiologists, some of the concerns include, but are not limited to, the following: aspiration risk, the use of neuromuscular blocking agents or opioids, cervical spine mobility, mouth opening, and the possibility of tracheal stenosis in patient with prior history of mechanical ventilation.

Case Report

A 33-year-old female with Type III spinal muscular atrophy presented for an urgent esophagogastroduodenoscopy (EGD) to evaluate for an upper gastrointestinal bleed with recent history of bloody emesis, melena, and abdominal pain. The patient was diagnosed with spinal muscular atrophy at the age 10. On presentation, the patient was wheelchair bound and was unable to lift her lower extremities against gravity, but she reported no upper extremity weakness or shortness of breath at rest.

Her past medical history was notable for a herniated cervical disk, hypertension, diabetes mellitus, fibromyalgia, cirrhosis, obstructive sleep apnea, asthma, current smoking, daily alcohol use and BMI of 34.4. Her medication list included insulin, propranolol, extended-release morphine, ondansetron, ceftriaxone, octreotide, and pantoprazole. The patient had a hysterectomy in 2014 and an EGD in 2018. Postoperatively, patient had some difficulty with breathing after emergence from anesthesia.

Her airway exam was notable for normal cervical spine range of motion, a Mallampati classification of III, normal mouth opening, and adequate thyromental distance. The neurologic exam was notable for weakness in her bilateral lower extremities and decreased range of motion at her hips and knees.

On the day of the procedure, the patient was brought to the endoscopy suite. Standard monitors consisting of non-invasive blood pressure, EKG, heart rate, pulse oximetry, and capnography were placed. After providing supplement oxygen via face mask, the patient underwent monitored anesthesia care with intravenous medications such as propofol titrated to effect. The patient tolerated the sedation and EGD well without airway compromise. At the end of the procedure, the patient underwent recovery in the post anesthesia care unit without complications. She was later discharged back to the inpatient medicine wards.

Discussion

Patients with SMA commonly require anesthesia to undergo diagnostic testing, procedure, or surgery. Airway and pulmonary complications are the most common presenting perioperative issues, especially in patients with SMA I and II.[5] Patients with SMA should have a thorough preoperative airway and respiratory evaluation. Patients with SMA often have limited mobility of the cervical spine secondary to contractures or prior surgeries, and patients with severe SMA can suffer

from reduced mouth opening due to ankylosis of the mandible.[5] Patients with a history of mechanical ventilation should be evaluated for possible tracheal stenosis.[5] In those with suspected difficult intubations, video laryngoscope, fiberoptic scope or laryngeal mask airways have been shown to increase success, and, in certain cases, retrograde intubation is an alternative to successfully establishing an airway. Tracheostomy or cricothyrotomy remain a back-up alternative to establishing an airway especially in emergencies.[5]

Patients with SMA are prone to gastroesophageal reflux, which affects nearly 100% of SMA type I patients, and most patients with SMA II and III report some reflux symptoms.[3] Patients with reflux should be appropriately evaluated and prophylaxis with antihistamines or proton pump inhibitors is recommended.

Postoperative respiratory planning is paramount in patients with SMA, especially in those with poor cough function or respiratory muscle weakness. Patients with normal cough function and strength are not at increased risk of postoperative respiratory complications.[6] Patients with SMA type I or II require hospital stays for respiratory support even after small procedures. One study conducted at a large center recommended oxygen titration to 95% saturation before extubating in patients with SMA types I and II.[5] Patients with SMA types III and IV that generally do not need nocturnal respiratory support most likely will not need respiratory support after surgeries; although, patients with SMA type III occasionally need support during acute illness or in severe disease.[5]

Anesthesia drugs have been used with varying success in patients with SMA, with neuromuscular blocking agents and opioids being the most important. A case review of 47 different case reports of patients with SMA, found no reported medication reactions are specific to SMA patients.[5] Nondepolarizing neuromuscular blocking agents have been shown to have prolonged effects even after reversal agent administration, and sensitivity in patients using nondepolarizing muscle relaxants appears to vary.[7] A large scale study on children with SMA concluded that children given neuromuscular blockade should be monitored closely with train-of-four stimulation as well as demonstration of muscle strength prior to extubation.[8] Patients with SMA should never be given succinylcholine due to its contraindication in neuromuscular disease, as these patients are at high risk for severe hyperkalemia.[6] Opioids have been used in perioperative care of patients with SMA, with the major concern being respiratory depression. Because of this, short-acting opioids are better suited for intra-operative use, along with careful titration and constant monitoring.[7]

Our patient underwent monitored anesthesia care for an urgent endoscopy without complications. During the case, opioids and neuromuscular blockade were avoided. The patient remained stable during the procedure and in the post-operative phase.

Conclusion

Patients with SMA often require anesthesia for a wide range of procedures and surgeries to improve their quality of life. Anesthetic management of these patients can be tricky, but with careful assessment and planning, anesthesia can be safely delivered to these patients. We presented the case of a young female with Spinal Muscular Atrophy who underwent monitored anesthesia care for evaluation of an upper gastrointestinal bleed. The most important anesthetic considerations for patients with SMA, include aspiration risk, the use of neuromuscular blocking agents and opioids, cervical spine mobility, mouth opening, and the possibility of tracheal stenosis in patient with prior history of mechanical ventilation.

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