



Managing a difficult airway in a patient with central core disease

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Introduction

- Two crucial roles of the anesthesiologist are to maintain the airway and to manage co-morbidities through the peri-operative period
- Optimizing the intubating conditions to aim for first pass success include adequate depth of anesthesia, complete neuromuscular relaxation, clear laryngoscopic view, and stable cardiopulmonary function, of which many co-morbidities can interfere with
- Succinylcholine is a widely used neuromuscular blocking agent characterized by a rapid onset and short duration and provides excellent intubation conditions.
- Succinylcholine, as well as volatile anesthetics, are known triggers of malignant hyperthermia and should be used with caution in at risk populations
- Central core disease is associated with an RYR1 mutation which increases risk for malignant hyperthermia and can make intraoperative management difficult for the anesthesiologist
- This case reflects how central core disease can present difficulty with airway management due to MH susceptibility, cranio-fascial abnormalities, and in this case morbid obesity

Case Report

- We present a 21-year-old male with a central nuclear myopathy, morbid obesity, and OSA presenting for cystoscopy for renal calculi.
- After careful preoperative evaluation, anesthetic plan was TIVA under LMA.
- The patient was induced with propofol, lidocaine, and fentanyl and initial attempt placing iGEL size 4 was unsuccessful due to inadequate mouth opening.
- An iGEL size 3 was placed with difficulty although the seal was inadequate at which point the patient was assisted back to spontaneous ventilation and awakened from anesthesia.
- As the urologist indicated the case was necessary to proceed, the decision was made to continue with an awake fiberoptic intubation.
- The patient's mouth was topicalized with 4% lidocaine and both an Ovasapyan and a Williams airway were attempted to be placed into patient's mouth, but the mouth opening was inadequate to accommodate the airways.
- Decision was then made to proceed with a nasal fiberoptic intubation.
- Afrin and 2% topical lidocaine were applied to each nares then serial nasal dilations were performed using progressively larger nasal trumpets (24 to 32 Fr)
- The fiberoptic scope was passed through the R nares to reach the glottic opening.
- A 6.0 micro laryngeal tube was passed through the vocal cords with some difficulty due to distorted anatomy and an omega shaped epiglottis.
- End tidal CO2 was confirmed and the patient was maintained on propofol infusion.



Figure 1: Patient with clinical features of central core disease⁴

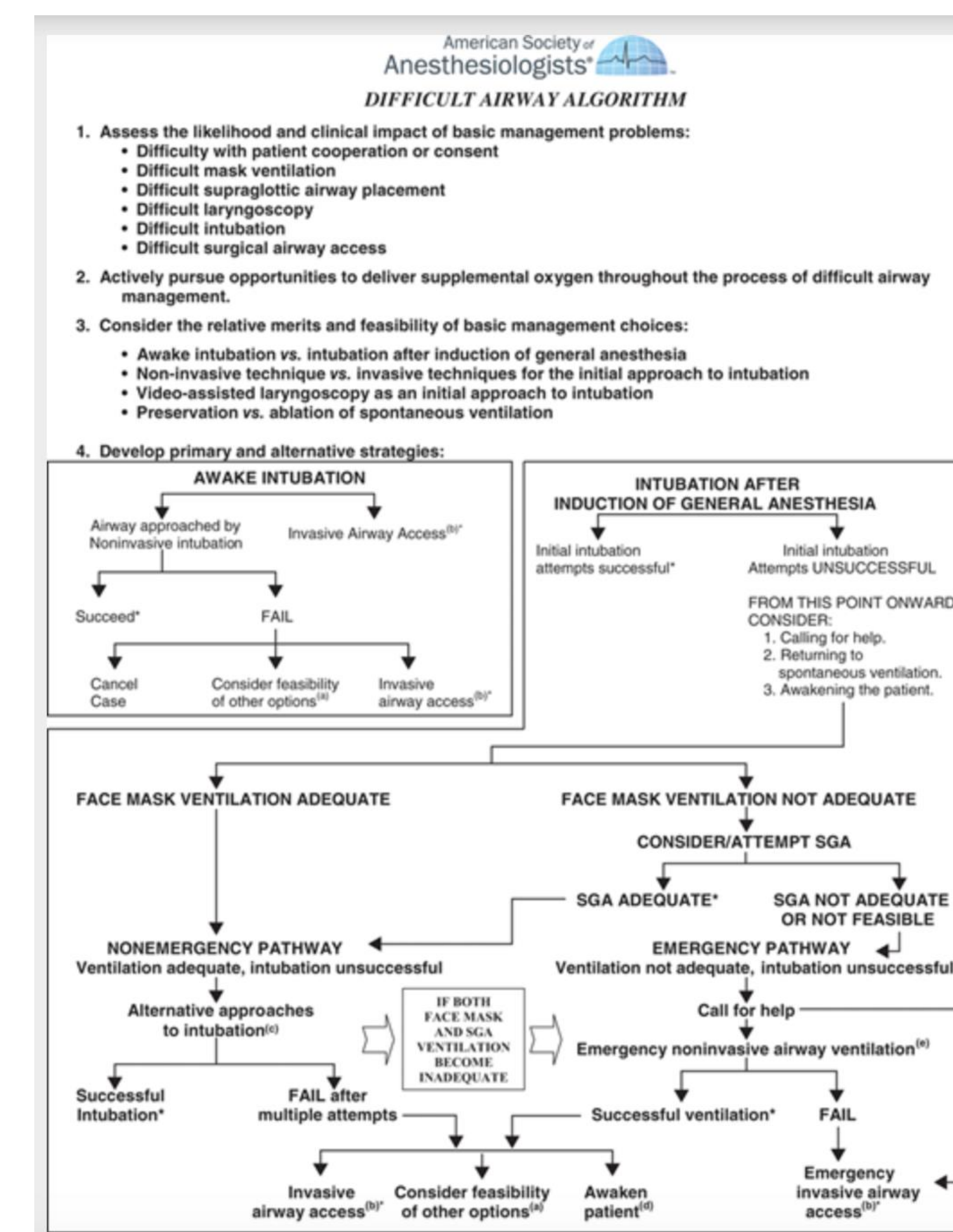


Figure 2: Difficult airway algorithm⁵

Discussion

- There are multiple congenital diseases that are associated with a higher risk of malignant hyperthermia, including central core disease
- For the anesthesiologist, anesthetic plans must be significantly altered when managing patients who are malignant hyperthermia susceptible
- Optimal intubating conditions are essential for anesthesiologists to achieve successful first-pass airway manipulation which can be difficult to achieve **without the use of neuromuscular blockade**
- Optimizing the intubating conditions to aim for first pass success include adequate depth of anesthesia, complete neuromuscular relaxation, clear laryngoscopic view, and stable cardiopulmonary function, of which many co-morbidities can interfere with
- Succinylcholine is a widely used neuromuscular blocking agent characterized by a rapid onset and short duration and provides excellent intubation conditions, although succinylcholine is contraindicated in central core myopathy due to risk for life-threatening hyperkalemia and malignant hyperthermia
- Central core myopathies involve mutations in the ryanodine (RYR1) receptor** making this patient population susceptible to MH
- Central core myopathy is also characterized craniofacial abnormalities that can cause airway management difficult, such as in with this patient and the use of depolarizing neuromuscular blockade to optimize intubating conditions is contraindicated making airway management even more challenging
- The difficult airway algorithm provides a structured framework for the anesthesiologist

Conclusion

- This case highlights the complex airway considerations patients with central core disease, a disorder associated with RYR-1 mutation and related malignant hyperthermia susceptibility, where avoidance of triggering agents, such as succinylcholine and volatile anesthetics, is lifesaving although limited use can be troublesome in the setting of advanced airway / failed airway management
- Although anesthetic plan was tailored appropriately to MH susceptibility, inadequate seal on the LMA reveals how myopathic patients, in addition to morbid obesity, may present with unexpected anatomical or functional airway challenges with limited resources of airway management
- Following the difficult airway algorithm, transition to awake fiberoptic was the next safest thing
- This case emphasizes the critical role of meticulous preoperative planning, the availability of non-triggering anesthetic options in patients with underlying myopathies and MH risk, anticipation of difficult airway scenarios, and execution of advanced airway management following the difficult airway algorithm

References

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