A Narcotic-free, Dexmedetomidine-based anesthetic for a child with Mitochondrial Myopathy

Irem Kaplan, MD1; Jan Wong, MD1,2
1SUNY Upstate Medical University, 2Upstate Golisano Children’s Hospital

INTRODUCTION
Mitochondrial disorders are a group of diseases with wide variability in presentation relating to degree of severity and organ system involvement.

Two major classifications of mitochondrial disorders: disruptions in the respiratory chain involved in oxidative phosphorylation to form ATP; and the other is related the metabolism of fatty acid as an alternative source of energy.

Mitochondrial diseases may involve either or both categories. This yields important anesthetic considerations such as potential for respiratory failure, cardiac depression/dysrhythmias, and exacerbation or new-onset muscular weakness.

CASE DESCRIPTION
This case involved a 16-year-old, 73-kg male with history of mitochondrial myopathy. Von Willebrand's disease, seizure disorder, gastrostomy feeding tube—dependence due to short bowel syndrome, and dystonia. The patient presented for a Broviac catheter placement for chemotherapy for acute myelogenous leukemia.

The patient’s mother reported that the patient had severe ileus after many previous anesthetics. This yields important narcotic use in several previous anesthetics.

ANESTHETIC MANAGEMENT
After premedication with midazolam 2mg IV, the patient was taken to the OR with an infusions of DDAVP and Dextrose 10% at 75ml/hr.

The patient was sedated with intravenous dexmedetomidine 1mg/kg bolus followed by maintenance infusion of dexmedetomidine at 0.8 mcg/kg/hour supplemented with oxygen and sevoflurane ranging from 0.4-2.2% through an classic LMA 4.

Spontaneous ventilation was maintained. Intra-operatively, the HR varied from 55-109beats/min. The BP was maintained at 104-144/36-84 mmHg. No abrupt changes in end-tidal carbon dioxide or oxygen saturation were noted.

The surgeon injected lidocaine into the incision site at the end of the procedure for pain control.

At the conclusion of the procedure, the LMA was removed and patient was taken to the PACU. He had no additional medication requirement and was quickly discharged from the PACU. Due to his medical complexity, the patient was observed overnight by the surgical team.

DISCUSSION
Our patient's post-operative course was uncomplicated, which was unique for this patient as he often had significant ileus and/or muscular weakness after previous anesthetics.

The strong history of post-operative ileus was likely exacerbated by opioid pain medications, which we were able to avoid with the use of dexmedetomidine.

Furthermore, avoiding propofol is beneficial in patients with mitochondrial disease as propofol is known to have the potential for significant muscular weakness.

There were no signs of cardiac or respiratory complications which are also important considerations in this patient population.

CONCLUSIONS
Dexmedetomidine can provide sufficient procedural sedation and analgesia for patient’s with mitochondrial disorders having central catheter placement. This is important as this population of patients often have worsening of muscular weakness with exposure to propofol.

Additionally, in patients with a strong history of ileus, the opioid-sparing effects of dexmedetomidine are of added benefit.

REFERENCES