Acute intermittent porphyria (AIP) can manifest as neurovisceral crises:
- Abdominal pain, dysautonomia, respiratory and limb weakness and seizures.

Triggers include many aspects of the perioperative setting:
- Fasting, stress and medications.

Thorough pre-operative preparation is essential:
- Consult up-to-date lists of safe medications in AIP.
- Minimize exposure to potential triggers.
- Make available medications to manage an AIP crisis.

Ultrasound-guided regional anesthesia can be performed safely.

Preoperative Management

A 9-year-old, 25.7 kg male with a confirmed genetic marker for acute intermittent porphyria (AIP) presented for right inguinal hernia repair. The family history was rife with anesthesia complications: one relative died on induction with a barbiturate, an aunt had an AIP episode while under anesthesia necessitating alkaline heme treatments, and the patient’s mother had “nearly died” during an anesthetic exposure.

Pre-operative management included a thorough AIP literature search, a metabolism and genetics consultation, and extensive communication with the surgeon and the patient’s mother prior to the surgery. Lists of safe medications in AIP were reviewed and a pharmacist was contacted. Gabapentin and alkaline heme were ordered and available in the pharmacy in case of a crisis.

The patient was encouraged to drink clear white grape juice until 2 hours prior to surgery. Per metabolism/genetics, the patient received an intravenous (IV) catheter in the holding area, and an infusion of dextrose 10%/saline 0.45% (D10/0.5NS) was started. A comprehensive metabolic profile was checked. In lieu of giving oral midazolam (due to conflicting safety data in AIP patients), we arranged a parental induction and gave IV midazolam (1mg/kg) and propofol 1mg/kg. We maintained the patient’s airway with a laryngeal mask airway (LMA) and administered sevoflurane. We performed an ultrasound-guided ilioinguinal-iliohypogastric nerve block with clear visualization of both nerves. Four mL of 0.25% bupivacaine and 1:200,000 epinephrine were injected with good spread throughout the ilioinguinal and internal oblique muscles. The D10/0.5NS infusion was continued; a glucose check was within normal range. A left inguinal hernia was discovered, necessitating the obtaining of surgical consent. Morphine 1.5mg IV was given prior to initiation of left-sided surgery. Ondansetron 4mg IV was administered during skin closure.

Intraoperative Management

We performed a parental IV induction with propofol, placed a laryngeal mask airway (LMA) and administered sevoflurane. We performed an ultrasound-guided ilioinguinal-iliohypogastric nerve block with clear visualization of both nerves. Four mL of 0.25% bupivacaine and 1:200,000 epinephrine were injected with good spread throughout the ilioinguinal and internal oblique muscles. The D10/0.5NS infusion was continued; a glucose check was within normal range. A left inguinal hernia was discovered, necessitating the obtaining of surgical consent. Morphine 1.5mg IV was given prior to initiation of left-sided surgery. Ondansetron 4mg IV was administered during skin closure.

Discussion

Acute intermittent porphyria (AIP) is an autosomal dominant deficiency in porphobilinogen deaminase that can manifest as a sudden, possibly fatal crisis, symptoms of which include severe abdominal pain, seizures, respiratory and limb weakness, hypertension, and tachycardia. Triggers include many aspects of the perioperative setting, such as fasting, stress and medications.

Our case provides evidence of the safe use of general and regional anesthesia in an AIP patient with an extensive family history of anesthesia complications. Thorough pre-operative preparation is essential for safe and effective care of AIP patients. Up-to-date lists of safe medications in AIP should be consulted. The likelihood of potential triggers during the perioperative period should be ameliorated. Medications to manage a crisis should be available.

References