Case Description
A 9-year-old Caucasian female with ADCY5-related dyskinesia, hypotonia, and motor delay were first noted at 6 months of age. She subsequently developed severe myoclonus, abnormal movements of the mouth, limbs, and trunk that increased in severity and frequency. She currently has nearly continuous painful choreiform movements that interfere with walking, speaking, and writing.

After multiple rounds of diagnostic testing, whole exome genetic sequencing identified a heterozygous R418W mutation in the ADCY5 gene.

The patient was admitted to our institution for deep brain stimulator placement as a treatment modality for her dyskinetic symptoms.

Clinical Course
The patient was admitted 24 hours prior to surgery for dextrose-containing IV fluid hydration to avoid entering a catabolic state while fasting. She received D10 + 0.2% NaCl + 20 mEq/L KCl at maintenance after she was made NPO 8 hours prior to surgery.

On the day of surgery, she received IV midazolam for premedication. General anesthesia was induced with lidocaine 1.5 mg/kg, fentanyl 1 mcg/kg, and propofol 4 mg/kg. The patient was easily bag-mask ventilated. Successful atraumatic oral intubation was performed.

General anesthesia was maintained with sevoflurane at approximately 0.8 MAC throughout the case, with no additional analgesia required. Ondansetron 1 mg/kg was given 30 minutes prior to extubation for PONV prophylaxis. Emergence, extubation, and a 1-hour PACU stay were uneventful.

Anesthetic Considerations
ADCY5-related dyskinesia is a rare movement disorder caused by a gain-of-function mutation in the ADCY5 gene resulting in increased cAMP production in tissues. Patients with ADCY5-related dyskinesia presenting for surgery offer several unique challenges for the anesthesiologist related to the pathophysiology of their disorder.

ADCY5-related dyskinesia is a rare movement disorder caused by a gain-of-function mutation in the ADCY5 gene resulting in increased cAMP production in tissues. Patients with ADCY5-related dyskinesia presenting for surgery offer several unique challenges for the anesthesiologist related to the pathophysiology of their disorder.

These include avoidance of exacerbating factors such as stress and β-adrenergic agonists, vigilant perioperative pain management to avoid excessive endogenous catecholamine release leading to exacerbation of symptoms, and consideration of multimodal analgesia as patients may have cellular tolerance to opioids.

Discussion
Thus far, only 15 individuals with ADCY5-related dyskinesia have been reported. We describe a female pediatric patient with ADCY5-related dyskinesia who successfully underwent general anesthesia at our institution. This is a novel case report since to date, no reports have been published describing the perioperative care of these patients.

Most patients with ADCY5-related dyskinesia report that their symptoms are exacerbated with anxiety and stress; accordingly, every effort should be made to minimize excessive metabolic stress; one measure is to admit the patient for preoperative IV hydration with dextrose-containing fluids. Triggering agents (β-adrenergic, D1 dopamine, or A2A adenosine receptor agonists, as well as phosphodiesterase inhibitors) should also be avoided whenever possible. Conversely, β-blockers may play a role in symptomatic relief, especially for the acute stress of the perioperative period.

Adequate postoperative pain management is especially important, and more challenging, in patients with ADCY5-related dyskinesia. Uncontrolled pain causes increased metabolic stress and catecholamine release, which can exacerbate dyskinetic symptoms. Multimodal analgesia should be considered if pain control with opioids is inadequate at doses otherwise appropriate for the patient.

References