Anesthesia in an 8-year old child with familial dysautonomia (Reilly-day syndrome)

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Abstract

Background: Familial dysautonomia is a rare genetic disorder with autosomal recessive inheritance that has been mainly reported in the Ashkenazi Jewish population. Autonomic dysfunction, tendency to severe hyperpyrexia, vomiting and recurrent pulmonary aspiration, and pain conception disorder, make a patient an interesting candidate for anesthesia.

Case Report: The patient was an 8-year old girl underwent surgery due to osteomyelitis and foot ulceration. Minimal drug, including sufentanil and nitrous oxide were used. Vital signs remained stable in the patient during the entire procedure.

Conclusion: Careful planning of the anesthetic management, preoperative hydration, understanding the physiological consequences, and being able to titrate the medications utilized are keys to decreasing complications encountered in these kinds of patients.

Keywords: familial dysautonomia, anesthesia, complications

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