Abstract # 204

Pregnancy in a Patient with Riley-Day Syndrome

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Familial dysautonomia (FD), or Riley-Day syndrome, is a rare congenital autosomal recessive neuropathy primarily seen in Ashkenazi Jews. Patients present with peripheral pain insensitivity, vomiting, aspiration, apnea, scoliosis and autonomic dysfunction. Intraoperative autonomic dysfunction can have significant implications on anesthetic management. Despite their insensitivity to pain, it appears essential to provide adequate analgesia to reduce surgical stress and dysautonomic crises.

A 22 yr old G1P0, at 35 wks gestation with FD was admitted for urgent cesarean section (CS) secondary to a 3 day history of fever, tachycardia, vomiting and subsequent fetal tachycardia. Her syndrome included pain insensitivity, self-mutilation, neurogenic bowel and bladder, renal failure, and autonomic dysfunction. She also had a history of Muscular Dystrophy (MD) and asthma, further complicating our anesthetic plan. After a multidisciplinary meeting and review of the literature, she underwent CS under general anesthesia with an arterial line and vigilant monitoring of urine output and temperature. Rapid sequence induction was performed with propofol and rocuronium. In the face of MD, succinylcholine and volatile anesthetics were avoided to decrease the risk of hyperkalemia and Malignant Hyperthermia. Anesthesia was maintained with a propofol infusion and intermittent fentanyl boluses. She was transferred intubated to the surgical ICU, where she was extubated 3 hours later. She was sent to the floor the following day and was discharged on POD 3.

Although parturients with FD and MD are at an increasingly high risk in the peripartum period, they can be managed safely with the careful planning of a multidisciplinary team. A thorough understanding of these disease states and their physiological consequences are imperative.

References: