A 24-yr male with Rubenstein-Taybi Syndrome (RTS) presented for ulnar collateral ligament repair. He had a history of developmental delay, communication impairment, ventricular septal defect closure, severe uncontrolled GERD, tracheomalacia, and anxiety. On exam, he had a narrow palate, dental crowding, cervical kyphosis, and adequate thyromental distance.

Our anesthetic plan was for standard monitors, awake fiberoptic intubation (AFOI), and a continuous supraclavicular nerve block for post-op pain. The patient received premedication of Bicitra, glycopyrrolate, fentanyl and midazolam, as well as home medications of pantoprazole and clonazepam. In the OR, a dexmedetomidine infusion was started (bolus of 1 mcg/kg, then continuous infusion at 1 mcg/kg/hr) and the airway topicalized with lidocaine. Aside from premedication, no further narcotics were administered. Prior to extubation, an ultrasound-guided supraclavicular catheter was placed.

Once extubated, the patient was transferred to PACU and connected to an OnQ pump with bupivacaine 0.125% at 6 ml/hr without bolus. He was discharged home from the PACU with pain well-controlled without additional medication. The catheter was removed 4 days post-op without complication and he transitioned successfully to oral Vicodin.

Discussion

Initially described in 1963, RTS is a complex genetic syndrome caused by a 16p13.3 disruption involving multiple organ systems and characterized by broad thumbs and toes, unique facial features, short stature, delayed language and mental retardation, that has significant anesthetic implications.

The combination of craniofacial and tracheal abnormalities, collapsible larynx, GERD, copious secretions, mental retardation, and risk of succinylcholine associated arrhythmias poses a significant challenge for intraoperative management. Given these concerns, we performed an awake fiberoptic intubation. Our patient was able to tolerate the AFOI very well, with dexmedetomidine allowing for a deep level of sedation without cardiovascular or respiratory side effects.

The supraclavicular catheter provided continuous analgesia without the need for difficult to interpret cues for pain and decrease narcotics to avoid further respiratory muscle impairment since RTS patients have significant airway collapsibility and high incidence of obstructive sleep apnea. The safety and efficacy of ultrasound-guided supraclavicular brachial plexus blocks under general anesthesia in pediatric populations has been established, as well as their placement under general anesthesia. The patient’s pain was well-controlled, not requiring oral pain adjuncts until the catheter was discontinued.

References