MCC-7016

Anesthetic Challenges of A Paediatric Patient with Rubinstein-Taybi Syndrome for Dental Day Surgery

Primary Author: Phui Sze Au Yong MBBS, MMed (Anaesthesiology)
KK Women's and Children's Hospital

Co-Authors: Evangeline Lim, ;

We describe a case of elective dental restoration for severe childhood caries in a 13 year old, 40kg, 1.5m Asian boy with Rubinstein-Taybi Syndrome with no previous anaesthetic history at our specialized paediatric unit. He had dysmorphism, global development delay, intellectual disability, severe myopia and surgically-closed patent ductus arteriosus. Cooperativity was limited even for his main caregiver, who gave a history of needing multiple adults to restrain him for eyewear fitting. Premedication worked with limited success to allow patient to tolerate an inhalational induction only initially. We describe our plans for an anticipated difficult airway to eventually achieve successful nasal intubation.

Multimodal analgesia allowed patient to wake up smoothly without emergence delirium. He was eventually monitored in general ward and discharged home uneventfully.

Discussion

Rubinstein-Taybi Syndrome is a rare autosomal-dominant syndrome with a paucity of literature on anesthetic management especially in Asian population. It poses anaesthetic challenges due to mainly airway, cardiac and skeletal anomalies, with wide interindividual variability. This is complicated in dental surgery by need for nasal intubation in a fretful, uncooperative child with potential difficult airway who is difficult to restrain due to his physical strength. A normal airway assessment does not preclude the possibility of a difficult airway. There are some reports of difficult mask ventilation and laryngoscopy. The patient had copious secretions which may predispose to risk of gastric aspiration. A potential pitfall was the lack of IV access before induction, and the risk of laryngospasm on IV cannulation as he was not premedicated with EMLA.

Another significant challenge was overcoming his stranger anxiety, which is contributed by previous bad experiences, severe myopia and an inability to verbalize. Communication, coordinating parental roles and premedication served important roles in coaxing the child into tolerating inhalational induction. We tried to avert forceful restraining techniques as that would have negative psychological impact and worsen the conduct of future anaesthetics.

In conclusion, perioperative planning and close multidisciplinary communication is essential to cater the anaesthetic to children with difficult airways. Careful consideration is needed to decide if this population is suitable as day cases, and anesthetic technique needs to be modified to include opioid-sparing analgesia in order to facilitate their smooth discharge as day cases because their pain may be much harder to manage at home.