A Novel Nasal PAP Mask Assembly Provided Continuous Active Oxygenation in a Patient with Fragile X Syndrome, Autism and Poor Face-Mask Fit during GA Induction for EUA and Dental Restoration

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Introduction: Fragile X Syndrome (FXS) is a rare disease characterized by intellectual disability and distinctive physical features including long and narrow face, large ears, a prominent jaw and forehead. These patients’s dysmorphic facial features may make poor face-mask fit and face mask ventilation difficult. A simple nasal PAP mask assembly has been shown to provide nasal CPAP to improve oxygenation in sedated obese patients with OSA and continuous active oxygenation in patients with difficult airway during GA induction and endotracheal intubation (1-5). We used this technique to improve ventilation in a FXS patient during GA induction.

Case Presentation: A 31-year-old FXS male with autism and poor dentition and multiple dental caries presented for examination under anesthesia and dental restoration. He was uncooperative with oral examination in the dentist’s office and airway examination in the anesthesia holding area. An infant face mask (size #2) with fully-inflated air cushion was shown to him and his parents. He was fitted with the mask over his nose and breathed through it comfortably. His parents gave their consent to use for taking photography and case presentation.

He became cooperative following 4 mg IV midazolam. He was then taken to the OR and pre-oxygenated with the infant mask secured over his nose. It was connected to a breathing circuit and the anesthesia machine with 4 LO2/min (4 cm H2O CPAP). Following GA induction with lidocaine/propofol, the nasal mask was used to deliver pressure-controlled ventilation (PIP 15 cm H2O, PEEP 5 cm H2O, RR 20/min). With continuous pressure-controlled nasal ventilation and oxygenation, oral endotracheal intubation was accomplished using video-laryngoscope without desaturation (Photo). He maintained 100% SpO2 throughout GA induction/intubation and tolerated the 6-hours procedure under GA without complication. Post-extubation, he required frequent oral suctioning with continuous nasal CPAP oxygenation. He recovered from anesthesia and was discharged home without delay.

Discussion: This nasal PAP mask assembly provided nasal CPAP pre-oxygenation and pressure-controlled nasal ventilation during GA induction and intubation in the uncooperative patient with Fragile X Syndrome, dysmorphic facial features and poor face-mask fit. It maintained continuous active oxygenation and allowed calm attempts to perform oral intubation using video-laryngoscope to avoid causing dental or oral injuries. It may improve patient safety at a low cost.
