INTRODUCTION

Pierre Robin Syndrome (PRS) associated with bilateral temporomandibular joint (TMJ) ankylosis and severe obstructive sleep apnoea (OSA) is a relatively uncommon occurrence in the paediatric age group. We discuss the peri-operative management of a child scheduled for bilateral mandibular distractor placement.

CASE REPORT

A 5-year-old girl weighing 15 kg, case of PRS with bilateral TMJ ankylosis presented with gradual reduction in mouth opening for 2½ years; ultimately, resulting in inability to open the mouth for the past 6 months. This was associated with feeding difficulty, disturbed sleep, excessive snoring and severe chest retractions while sleeping.

Airway examination revealed a nil mouth opening with retro-micrognathia [Figure 1a and b]. Neck movements remained unaffected. Lateral cephalogram showed a narrowing of posterior pharyngeal space [Figure 2a]. Computed tomography three-dimensional face was suggestive of the retro-micrognathic mandible with bilateral TMJ ankylosis, partial fusion of vertebral body of C2–C3, posterior elements of C2–C3 and C5–C6 along with glossoptosis causing mild compression of nasopharynx and oropharynx [Figure 2b]. Polysomnography showed apnoea + hypopnoea (A + H) episodes: 36.7/h, average oxygen saturation: 93%, oxygen desaturation events:
32.0/h, lowest oxygen saturation: 63% and Respiratory Distress Index: 36.7%; suggesting severe OSA. Two-dimensional echo imaging was unremarkable.

In view of the anticipated difficult airway, high-risk consent was taken, and fibreoptic nasotracheal intubation planned, with Ear, Nose and Throat team standby for tracheostomy.

The child was kept nil orally on the morning of the planned surgery as per standard paediatric fasting guidelines. Pre-operative preparation included: (a) Securing intravenous access pre-emptively using eutectic mixture of local anaesthetics (EMLA) cream (b) Injection glycopyrrolate 4 µg/kg IV (c) nebulisation with 2 ml of 4% lignocaine (d) Injection dexamethasone 4 mg IV (e) Injection ketamine 5 mg IV (f) decongestion of the nasal mucosa of both nostrils using 1–2 drops of 0.05% xylometazoline nasal drops to prevent bleeding. In addition, the right nostril was packed with pledgets soaked in 2 ml of 2% lignocaine with 5 µg/ml adrenaline. The child was subsequently wheeled into the operation theatre and monitors in accordance with minimum monitoring standards, plus the Masimo Root Monitoring System (Root®) for Oxygen Reserve Index (ORI) monitoring was attached.

Inhalational induction using 100% oxygen @ 5 l/min with incremental sevoflurane (2%–4%) was done. Simultaneously, an intravenous bolus of 1 µg/kg dexmedetomidine was given over 10 min, following which infusion @ 0.5 µg/kg/h was started. On attaining sufficient depth of anaesthesia, a shortened, well-lubricated 4.5 mm ID uncuffed endotracheal tube was gently inserted into the left nostril and closed breathing circuit attached to it to deliver oxygen-sevoflurane mixture while maintaining spontaneous ventilation.

A 2.8 mm outer diameter paediatric fibroscope was used to perform flexible fibreoptic intubation using a well-lubricated 4.5 mm Halyard™ microcuff endotracheal tube and its placement was confirmed by capnography. Flow-volume loops were checked for any air leaks and the micro-cuff was inflated to maintain a cuff pressure of 10 cm H₂O. Subsequently, fentanyl 2 µg/kg and atracurium 0.5 mg/kg was administered and mechanical ventilation initiated.

The patient had partial airway obstruction with subcostal and intercostal chest retractions during inhalational induction. However, it was not accompanied by oxygen desaturation or bradycardia. Pre-induction ORI value was 0.35, which reduced to 0.29 by the time the improvised nasopharyngeal airway (NPA) was inserted to deliver oxygen-sevoflurane mixture. With the NPA in-situ and spontaneous ventilation being maintained, ORI value steadily rose to 0.46, following which flexible fibreoptic intubation was attempted. It further dropped to 0.1 by the time of endotracheal tube placement and confirmation, although without bradycardia or oxygen desaturation. ORI monitoring was discontinued following intubation.

Anaesthesia was maintained using oxygen, air and desflurane, titrated to keep a Minimum Alveolar Concentration of 1–1.2 using low flow anaesthesia plus dexmedetomidine infusion @ 0.5 µg/kg/h with intermittent boluses of atracurium. The surgery
lasted three and a half hours and the intra-operative period remained uneventful. The child was electively ventilated in view of the anticipated difficulty in maintaining patency of the airway following on-table extubation. Post-operatively, the child was sedated using 1 µg/kg/h fentanyl and propofol infusion @ 75 µg/kg/min, which were stopped the next morning. Once fully awake, a spontaneous breathing trial was given, and the child was uneventfully extubated. The child was subsequently put on heated, humidified high flow nasal cannula (HHHFNC) oxygen therapy (using AIRVO™ 2 Humidification System) for 24 h and sips of clear fluids were started in the evening. The child had a good recovery, and graded mandibular retraction was started from the 3rd post-operative day onwards.

**DISCUSSION**

PRS is a sequence of events occurring as a result of the triad of micro/retrognathia, glossoptosis and airway obstruction. The severity of PRS and the associated symptoms usually improve with age; however, our patient became more symptomatic and the airway obstruction worsened due to the development of bilateral TMJ ankylosis. TMJ ankylosis can be classified as congenital, idiopathic, traumatic or infective. Our patient had a forceps delivery and history of perinatal trauma, suggestive of a possible traumatic aetiology. Association of PRS and TMJ ankylosis increases the likelihood of sleep-disordered breathing disorders such as OSA due to the hypoplastic and micrognathic mandible. OSA complicates anaesthesia management as it causes significant cardiac, pulmonary and central nervous system impairment because of chronic oxygen desaturation leading to pulmonary hypertension and eventually cor pulmonale. Thus, diagnosing OSA and optimising the child is of paramount importance. Apart from eliciting a thorough history, polysomnography remains the “gold standard” for diagnosing OSA.

No case of PRS with bilateral TMJ ankylosis has been reported till date. Taking into perspective the relevant anaesthetic concerns and patient safety, we incorporated ORI monitoring at induction and use of HHHFNC oxygen therapy in the immediate post-extubation period, in addition to performing flexible fibreoptic guided intubation.

ORI is a novel pulse oximeter-based non-dimensional index with a unit-less scale that ranges from 1.00 to 0.00 as PaO₂ decreases from 200 to 80 mm Hg. It is a non-invasive and continuous parameter which provides insight into the patient’s oxygen status in moderate hyperoxic range (PaO₂ >100 and <200 mm Hg), which is described as the patient’s oxygen “reserve.” This is a great advantage over pulse oximetry, which cannot herald an oxygen desaturation event until PaO₂ drops to 80 mm Hg. Thus, in clinical situations such as “securing a difficult airway,” ORI may prove to be a beneficial tool in estimating the time available at our disposal (before desaturation) for intubation. HHHFNC oxygen therapy in children is defined as heated, humidified and blended air/oxygen delivered through nasal cannula at different flow rates ≥2 l/min, delivering both high concentrations of oxygen and potentially continuous distending pressure. Suggested mechanisms of action include washout of nasopharyngeal dead space, reduction in inspiratory resistance and work of breathing, improvement of airway conductance and pulmonary compliance, and providing an end-distending pressure to the lungs. HHHFNC oxygen therapy in children with OSA reduces respiratory events, improves oxygenation and may also be effective in continuous positive airway pressure intolerant children.

**CONCLUSION**

Successful anaesthesia management of PRS with TMJ ankylosis and severe OSA involves thorough pre-operative evaluation and risk stratification, formulating a comprehensive airway management strategy for the peri-operative period, including preparation for extubation and post-extubation monitoring. Advances in anaesthesia monitoring and the use of sophisticated equipment have revolutionised peri-operative care, especially in challenging difficult airway scenarios, thereby significantly reducing the risk of potentially disastrous events.

**Declaration of patient consent**

The authors certify that the parents have given consent for their child’s images and other clinical information to be reported in the journal.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.
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