

Subglottic stenosis in an adolescent with pierre robin sequence: Expect the unexpected

Dear Editor,

A 17-year-old male (weight-37 kg, height-152 cm), with a known case of Pierre Robin Sequence (PRS), was scheduled for mandibular distraction osteogenesis. He had a history of snoring and progressive feeding difficulty. On airway examination, Mallampati grade-IV with mouth opening of one finger, Chin, Oral opening, Previous Intubation, Uvula, and Range (COPUR) score 12, prominent buck teeth [Figure 1a and b]. Awake fiberoptic intubation was planned to secure the airway with tracheostomy back-up. The patient was prepared with 4% lignocaine nebulization, injection glycopyrrolate 4 μ g/kg, and appropriate counseling. Since the patient was anxious, dexmedetomidine was infused (1 mcg/kg followed by 0.5 mcg/kg/hour). Fiberscope was inserted with flexometallic tube (FMT)-6.0 mm in situ. Subglottic stenosis was seen during Fiberoptic Intubation (FOI) [Figure 1c], however, successful intubation was expected as a smaller size FMT was used compared to the age of the patient. But Endotracheal Tube (ETT) could not be negotiated even after maneuvering. Due to repeated attempts, the procedure became traumatic. The patient started to cough vigorously, became restless and combative. Suddenly, he desaturated to 88% so bag mask ventilation (BMV) was attempted but fare amount of resistance was encountered. Thus modified rapid sequence induction (RSI) performed with IV fentanyl-2 mcg/kg, propofol-2 mg/kg an succinylcholine-1.5 mg/kg. Following which compliance of BMV improved. Simultaneously tracheostomy was also performed and a tracheostomy tube-6.0 was placed [Figure 1d]. Anesthesia was maintained with O₂:air-50:50 with sevoflurane and intermittent vecuronium. Along side IV hydrocortisone-50 mg, etofylline-35 mg, theophylline-12 mg, epinephrine (1:1000)-0.3 mg intramuscularly, 8–10 puff of salbutamol and ipratropium bromide was given to relieve bronchospasm. Intra-operative course was uneventful. After successful completion of the surgery, the patient was shifted to ICU for gradual weaning. The tracheostomy tube was removed on the fourth post-operative day (POD) and discharged from the hospital on POD10th. The patient was kept on follow up for the next one month without any further complications.

Pierre Robin Sequence is a combination of mandibular hypoplasia, postero-superior displacement of tongue and cleft palate.^[1] The incidence of PRS varies from 1:5000 to



Figure 1: (a) and (b). Anterior and lateral view showing micrognathia, limited mouth opening and buck tooth. (c). Subglottic stenosis during fiberoptic intubation. (d). Postoperative period with tracheostomy tube in situ

1:85,000.^[1,2] The gold standard technique used for difficult airway management in PRS is FOI.^[3] Other available techniques include Glidescope, Airtraq, Air-Q scope, Shikani scope, laryngeal mask airway, retrograde technique, etc.^[1] The incidence of subglottic stenosis varies from 0 to 2% in neonates.^[4] Knapp *et al.*^[5] performed a retrospective review to conclude an increased incidence of subglottic stenosis in PRS and the requirement of a smaller size endotracheal tube.

In this case, FOI was performed using dexmedetomidine for conscious sedation. The procedure went smoothly till the visualization of the vocal cord. Because of the presence of the subglottic stenosis rail roading of the ETT became traumatic, which blurred the view. Subsequent attempts to place a smaller size ETT became difficult. Despite a thorough suctioning some amount of blood trickled into the trachea leading to bronchospasm and desaturation, which was managed uneventfully. Anesthesia plane deepened by modified RSI for smooth BMV and tracheostomy simultaneously. The patient was shifted to ICU for smooth removal of the tracheostomy tube. Further post-operative course was uneventful. To conclude, subglottic stenosis should be anticipated with adequate airway preparation while managing a case of PRS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients

understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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