

## Pierre robin sequence with cervicothoracic kyphoscoliosis: An anesthetic challenge

Pierre Robin sequence (PRS) is a congenital syndrome consisting of micrognathia, receding and short mandible, and cleft lip/palate. The major problem on PRS as faced by anesthesiologist is maintenance of airway in view of resultant glossoptosis.<sup>[1]</sup>

A 14-year-old male patient was posted for release of tethered spinal cord. He was operated for myelomeningocele at the age of 1 year. He had severe deformities of face, spine, and lower limbs, with high arched cleft palate, hypoplastic mandible, and short restricted movements of neck. The thorax was asymmetrical with dorsolumbar kyphoscoliosis of cervical and thoracic region. X-ray posteroanterior (PA) showed lateral curvature of the spine. Neck X-ray showed deviation of trachea, and narrowing of airway below the glottic region. He had difficulty to sit and was supported by two pillows while lying. On admission, his pulse rate was 88/min, BP-110/80 mmHg. His parents were explained the risk involved in placing the endotracheal tube and an informed consent was procured. The patient was taken to the operating room and O<sub>2</sub> was given by venturi mask after nebulizing with 2% lignocaine for 20 min. Intravenous (IV) line was secured and RL (Ringers Lactate) started. IV dexamethasone 2 mg and dexmedetomidine 25 microgram was given along with 100% O<sub>2</sub> for 5 min, subsequently, O<sub>2</sub> and sevoflurane were administered for 10 min. Later O<sub>2</sub> and sevoflurane were continued through nasopharyngeal airway attached via an universal 15 mm connector to breathing circuit. Fiberoptic intubation was done in presence of O<sub>2</sub> and

sevoflurane with great difficulty in negotiating the Endotracheal tube (ETT) as the glottic and trachea were deviated. The spinal surgery was uneventful in the prone position. He was electively ventilated overnight in intensive care unit (ICU) due to airway edema, and extubated on day 2. He was shifted from ICU after 4 days and discharged from hospital after 8 days.

Craniofacial abnormalities are commonly seen in PRS. In our case the child had all the classical signs of PRS, so a difficult airway was anticipated.<sup>[2]</sup> Oral premedication is usually given to reduce perioperative anxiety, ease induction, and increase parental satisfaction.<sup>[3]</sup> We chose not to sedate preoperatively this patient due to craniofacial deformity, cervicothoracic kyphoscoliosis, and PRS, instead administered titrated dose of dexmedetomidine in operation theatre, under standard monitoring. We even anticipated difficult percutaneous tracheostomy in case of emergency in this patient.

Patients with scoliosis suffer from restrictive lung disease which decreases vital capacity, functional residual capacity, tidal volume, and increased respiratory rate.<sup>[4]</sup> The severity of the deformity is best determined by measuring Cobb's angle. Numerous studies have documented that the more severe the thoracic curve (greater Cobb's angle) the more profound the disturbance on pulmonary function.<sup>[5]</sup> The preoperative assessment should focus on any cardiovascular, respiratory, or neurological impairment in patients with Cobb's angle >60°. Cardiology evaluation to assess ventricular size and evidence of pulmonary hypertension are required. Thoracic scoliosis causes a significant reduction in the number of alveoli predisposing these patients to impairment of gas exchange and pulmonary hypertension. These patients could present problems during extubation, with difficulty in weaning off the ventilator. Lilius also described associated malformations in 21.8% of cleft lip/palate children.<sup>[6]</sup> Hypoxia, hypercapnia,

acidosis and any increase in pulmonary vascular resistance should be avoided. Pulmonary function test and ABG are crucial and guide decisions regarding mode and time delivery and requirement of ventilator support.<sup>[7]</sup> Anesthesia poses a significant risk in this case as there is no single regimen that can be recommended for anesthesia management. The titrated dose of IV sedation with inhalational agent without muscle relaxant may be appropriate alongwith individualized perioperative care.

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### Conflicts of interest

There are no conflicts of interest.

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