

## CASE REPORT

# Effective intubation technique to manage the airway obstruction in A case of Pierre Robin sequence: A case report

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**Abstract**

The case was a 4-day-old boy referred to the hospital with a diagnosis of Pierre Robin Sequence and with airway Obstruction. Using a laryngoscope in the hyperextended position and putting pressure on the cricoid, the vocal cords were visualized, and with the help of an assistant, the intubation was performed.

**KEYWORDS**

airway obstruction, intubation, Pierre Robin sequence

## 1 | INTRODUCTION

Pierre Robin Sequence (PRS) is characterized by a sequence of events including mandibular hypotrophy (micrognathia), abnormal posterior placement of tongue (glossoptosis), and airway obstruction.<sup>1</sup> A wide U-shaped cleft palate is reported in most of the cases as well.<sup>2</sup> Pierre Robin sequence happens from 1/3120 to 1/14,000 live births.<sup>3,4</sup> The incidence rate of the PRS is equal in boy and girl infants.<sup>5</sup> The mortality rate of infants with RPS is reported to be between 3.6% and 21%.<sup>6</sup> It was shown that the SOX9 gene, an important chondrogenic regulator, has been associated with the occurrence of PRS.<sup>7</sup> Manifestations, severity, and functional situation are extremely variable in PRS cases, but many of them experience airway obstruction and feeding difficulties as the main manifestations of PRS. Airway obstruction can happen due to abnormal tongue positioning, which blocks the nasal and oral pharynx on inhalation.<sup>1</sup> The obstruction of the airway in PRS infants may be at the tongue base level, which can lead to apnea, frequent oxygen desaturations, and cyanosis. A high amount of energy is used for these

obstructive forces to continue breathing.<sup>8</sup> Feeding complications are believed to be a result of airway obstruction and cleft palate because they lead to insufficient negative intraoral pressure to extract the milk from the breast or the bottle.<sup>9</sup> Gastroesophageal reflux is a prevalent outcome of this procedure.<sup>10</sup>

Prone positioning and nasopharyngeal airway placement may assist to advance sufficient respiratory function, although long endotracheal intubation and surgical airway intervention may be required for severe cases.<sup>11</sup>

In this study, we describe a case of Pierre robin sequence with complicated efforts to manage the airway obstruction and using an intubation technique to make the endotracheal tube placement possible.

## 2 | CASE PRESENTATION

A 4-day-old, 3200-g boy, who was born at 36 weeks gestational age by cesarean delivery due to polyhydramnios and fetal distress, with an Apgar score of 9–10, was referred to hospital with a diagnosis of Pierre Robin Sequence based

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on micrognathia and glossoptosis. The patient was admitted with symptoms of tachypnea and a heart murmur. In the previous center, he was treated with an oxygen hood. There was no family history of congenital disorders. The mother was G4P2Ab1, with a history of hypertension and no history of diabetes.

The result of the primary arterial blood gas (ABG) test was as follows: PH: 7.10, Pco<sub>2</sub>: 89 mmHg, Po<sub>2</sub>: 36 mmHg, O<sub>2</sub> sat: 46%, and Hco<sub>3</sub>: 27 meq/L. The level of C-reactive protein (CRP) was 2 mg, and the levels of white blood cell (WBC) were  $13.6 \times 10^9/L$ , hemoglobin was 14 g/L, and platelet was  $281 \times 10^9/L$ . The result of echocardiography showed good left ventricular ejection fraction (LVEF), mild peripheral pulmonary stenosis (PPS), and patent foramen ovale (PFO).

On admission, oxygen saturation was 92%, and heart rate was 145. According to the ABG test, the patient had respiratory acidosis. Patients had frequent oxygen saturation drops. The prone position was not relieved the airway obstruction because of cleft palate and retraction of the tongue. Moreover, fixing the tongue with an endotracheal tube was not effective as well. Therefore, the patient needed intubation. The attempt of the pediatrician and anesthesia team for tracheal intubation was failed. Although the vocal cords were visible hardly by laryngoscope, they disappeared immediately and intubation was impossible. Finally, by using a laryngoscope, while the patient was in the hyperextended position and the physician at the sitting posture, by putting pressure on the cricoid, the vocal cords were visualized, and with the help of an assistant, the intubation was performed (Video S1). The laryngoscope used in this study was HEINE laryngoscope blade No. 1, and the diameter of the intubation tube was 3.5 mm. This intubation technique was successful at the first attempt. Ampicillin and amikacin antibiotics were primarily administered for these patients and then they changed to meropenem and vancomycin.

### 3 | DISCUSSION

Pierre Robin sequence usually leads to upper airway obstruction as a result of micrognathia, glossoptosis, and tongue displacement, which is contacted with the pharyngeal wall.<sup>12</sup> Positioning and nasal pharyngeal airway placement are the primary management for airway obstructions in PRS patients<sup>12</sup>; however, they may need surgical interventions such as mandibular distraction osteogenesis (MDO), tongue lip adhesion, tracheostomy, and subperiosteal release of the floor of the mouth.<sup>13</sup> Intubation in infants is a very challenging situation, and desaturation can happen very rapidly. Hard intubation associated with PRS may lead to cardiac arrest.<sup>14</sup> Therefore,

it is essential to develop techniques to manage difficult intubation in infants. In this study, patient was situated in the hyperextended position while the physician was in the sitting posture, by using a laryngoscope and putting pressure on the cricoid, the vocal cords have appeared, and the intubation was done with the help of an assistant.

Various techniques can be used for intubation. Takashita et al. used fiberoptic nasotracheal intubation to reduce the severe obstruction of the upper airway in PRS patients.<sup>12</sup> Zhang et al. used finger guide intubation to reduce severe upper airway obstruction in 2 PRS infants.<sup>15</sup> Sonwane et al. performed paraglossal intubation with Miller's blade 1 to manage the PRS airway obstructions and it was successful.<sup>16</sup> In Marson et al.'s study on 33 neonates, 37% of the cases were managed with endotracheal intubation, and 63% were managed with fiberoptic bronchoscope.<sup>11</sup> In Hayat Khan et al.'s study, successful intubation was done with the help of air-Q intubating laryngeal mask airway (ILMA).<sup>17</sup> In Agrawal et al.'s study, paraglossal straight blade intubation technique was used in 5 PRS patients to manage the airway obstruction.<sup>18</sup> In Semjen et al.'s study, the paraglossal approach joint with a gum elastic bougie made intubation easy for most patients with severe PRS. Thus, they suggested it as the first line for airway management in these patients.<sup>19</sup> In Runyan et al.'s study on 171 infants with PRS, nonsurgical [conservative] and surgical (tracheostomy or mandibular distraction osteogenesis [MDO]) were used to manage the airway obstruction. 11% of infants treated with conservative technique had an obstructive index (OI) >20, while 67.5% of those treated by MDO and tracheostomy met this severity measure. Tracheostomy was related to neurologic impairment as well as low birth weight among those treated by surgery, in comparison with the MDO group. Infants treated by conservative technique and with MDO had developed long-term feeding and airway obstruction results in comparison with the tracheostomy group.<sup>20</sup>

Allen et al. reported a PRS patient with obstructive airway signs as a result of the right-sided soft palatal mass comprising heterotopic neural tissue. Trans-palatal and trans-oral methods were performed for the removal of right-sided soft palatal mass, and the symptoms resolved after removal.<sup>21</sup>

### 4 | CONCLUSIONS

The case of this study was a neonate with airway obstruction associated with RPS that prone positioning, fixing the tongue, and several tracheal intubation attempts were failed to manage the obstruction. We conclude that by using a laryngoscope in the appropriate position and putting pressure on the cricoid, the vocal cords will

be visualized and intubation becomes possible. This improves the success rate of intubation in infants with severe PRS and can be suggested for airway management in this situation.

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## CONFLICT OF INTEREST

The authors declare no conflict of interest.

## AUTHOR CONTRIBUTIONS

SM and RF were involved with the clinical management of the patient and contributed to the preparation of this manuscript.

## CONSENT

The study is approved by the ethics committee of Mazandaran University of Medical Sciences. Written informed consent was obtained from the patient's parents.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author.

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## SUPPORTING INFORMATION

Additional supporting information may be found in the online version of the article at the publisher's website.

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