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CASE REPORT

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A case of Pierre Robin syndrome in a child with no soft palate and complications from pneumonia in Bangladesh

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Key Clinical Message

Children with Pierre Robin syndrome (PRS) often have trouble breathing and eating as soon as they are born. If conservative therapy fails to alleviate airway obstruction, surgical surgery may be considered. Patients with PRS require multidisciplinary approaches for treatment.

Abstract

Pierre Robin syndrome is a common craniofacial abnormality that causes glossoptosis and blockage of the upper airway. This renders it difficult to feed, which leads to severe malnutrition. This condition is also often marked by an absence of a soft palate. We mention a newborn with Pierre Robin syndrome with the absence of a soft palate and pneumonia complications, whose impending respiratory failure was treated successfully. To solve the complex problems that these babies and their families are facing, a multidisciplinary approach is needed.

KEYWORDS

congenital defects, feeding problem, micrognathia, pediatrics, Pierre Robin syndrome, pneumonia, soft palate

1 | INTRODUCTION

In 2004, WHO estimates that about 260,000 deaths worldwide were caused by congenital anomalies.[1] Every 4.5 min, a newborn is born with a birth defect.[2] Although the worldwide incidence of birth defects is estimated at 3%–7%, the rate varies widely between countries.[3] Pierre Robin syndrome (PRS) is a congenital disorder characterized by micro or retrognathia, breathing problems, feeding problems, cleft palate, and glossoptosis.[4,5] The incidence of this condition is uncertain but is estimated to occur in 1/8500–1/20,000 births.[5] Some arguments suggest an embryonic pathology in the development of caudal hind brain.[4] PRS is a clinically well-defined subgroup of the cleft lip-palate population with an unknown etiology, often observed as a part of other Mendelian syndromes, such as Stickler's syndrome, velocardiofacial syndrome, and Marshall's syndrome.[6]

Recent studies on genetics have shown that the association of dysregulation of the genes SOX9 and KCNJ2 may

be involved in PRS, as evidenced by a familial translocation with a breakpoint located in the gene empty region between SOX9 and KCNJ2, and by reduced expression of SOX9 and KCNJ2 in non-translocated patients with PRS.[7] Airway obstruction in PRS has been classically described as developing soon after birth.[8] However, this view was challenged by Ogborn and Pemberton, who reported that 5 of 16 (30%) of their cases presented with upper airway obstruction between 2 and 21 days of age.[9] Bull et al. noted that upper airway obstruction may appear over the first month of life; specific clinical details were not provided.[10] The late onset of upper airway obstruction, particularly after initial hospital discharge, has potentially serious consequences. In this case study, a newborn with Pierre Robin syndrome and an absence of soft palate was diagnosed after birth, and pneumonia complications later developed.

2 | CASE HISTORY

On last week of January 2023, a 1-day-old male neonate weighing 2500 g delivered via caesarian section presented to the pediatric emergency department because he had been breathing quickly and having difficulty feeding since birth. He also had been breathing loudly and having his chest stuck out since birth. After what seemed to be a normal prenatal period, she gave birth to him normally at the hospital when she was 36 weeks pregnant with her second child. Her brother was born through a normal vaginal delivery, but he died in the PICU within 7 days due to respiratory distress. Her second child cried as soon as he was born, but after 6 h of birth, he started breathing quickly and extremely difficult. Since then, his breathing has become loud. Since the newborn was born, the front of the chest wall stood out. The newborn had trouble eating from the time it was born and never learned how to suckle properly. A cup and spoon were used to feed him his mother's milk. He was taken to a pediatrician, who diagnosed he have a soft palate absent, micrognathia, glossoptosis, Transient Tachypnea of the Newborn (TTN), among other things. On examination, the newborn looked sick and was irritable. He had stridor in both phases of breathing and tachypnea (respiratory rate of 66/min), and there were clear retractions of the suprasternal, intercostal, and subcostal muscles. No cyanosis happened. The color of the newborn face was fine, and he had the Pierre Robin syndrome deformity (Figure 1). Then, he was admitted to the NICU of the Rupgonj, Narayangonj, Medical College Hospital, where pertinent blood investigations (Table 1), Chest X-ray (Figure 2), treatments (Table 2), and follow-up (Table 3) were received in accordance with the protocol. Pulse oximetry showed an oxygen saturation of 92%–94% in all four limbs in room air. Examination of

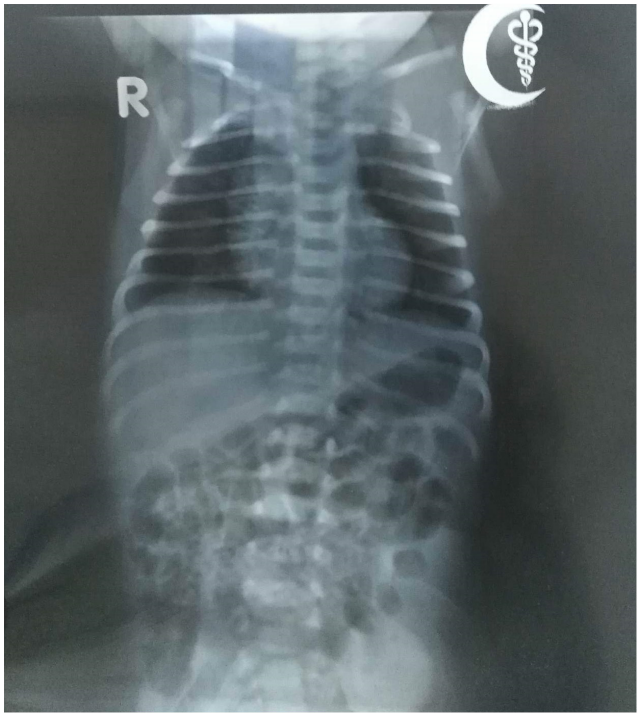

FIGURE 1 Chest X-ray of the patients.

the cardiovascular system was normal. He was, however, discharged on request (DOR) on Day 5 after birth, as a result of a financial issue, and was re-admitted to the government Maternal and Child Health Training Institute on next day, at 10.52 AM and diagnosed with pneumonia and Pierre Robin syndrome. He was receiving regular oxygen treatments, fluids, and antibiotics (Table 3). His bilirubin level increased to 15.1 on Day 9 after birth, at which point he was diagnosed with neonatal jaundice and began phototherapy. The next day, February 8, he was discharged with instructions to visit the pediatric surgery department. He was last followed up on by phone on 3rd week of February 2023, and he was now relatively stable in condition, and he will go for pediatric surgery consultation next month.

3 | DISCUSSION

Children with Pierre Robin syndrome often have trouble breathing and eating as soon as they are born. Due to microretrognathia and glossoptosis, which are physical problems with the airways, the large tongue falls back when the person is lying down.[11] This causes upper airway obstruction, which stops normal breathing and leads to low oxygen levels.[12]

Because they have trouble eating, the children do not gain much weight and show signs of aspiration pneumonia. Babies born too early may have a transitional

TABLE 1 Investigation parameters of the case.

Sl No	Name of the test	Date	Findings	Normal values
01	ABO type	30.01.2023	A (+ve)	
02	CXR of lung	31.01.2023	Bronchopneumonia	
03	S. Creatinine	31.01.2023	0.75 mg/dL	05–1.10 mg/mL
04	C-reactive protein (CRP)	31.01.2023	4.0 mg/L	Less than 10 mg/L
05	S. Sodium (Na)—mmol/L	31.01.2023	141	135–145 mmol/L
06	S. Potassium (k)—mmol/L	31.01.2023	4.8	3.5–5.0 (mmol/L)
07	S. Chloride (Cl)—mmol/L	31.01.2023	107	96–106 (mmol/L)
08	S. Calcium (Ca)—mmol/L	31.01.2023	8.3 mg/dL	8.6–10.3 mmol/L
09	Arterial blood gas test (ABG)	30.01.2023	PH: 7.43, pCO2-31.9, pO2-150, HCO3-21.4	
10	Hemoglobin (Hb) (g/dL)	31.01.2023	15.2	14.0–17.5 g/dL
11	White blood cells (WBC) (/L)	31.01.2023	14,000/cumm	4000–11,000/cumm
12	Platelets	31.01.2023	280,000/cumm	150,000–450,000/cumm
13	Neutrophil %	31.01.2023	68%	25%–66%
14	Mean corpuscular volume (MCV) (FL)	31.01.2023	107.1 fL	76–96 fL
15	S. Bilirubin	07.02.2023	15.1 mg/mL	Neonatal: 1.5–12.0 mg/mL

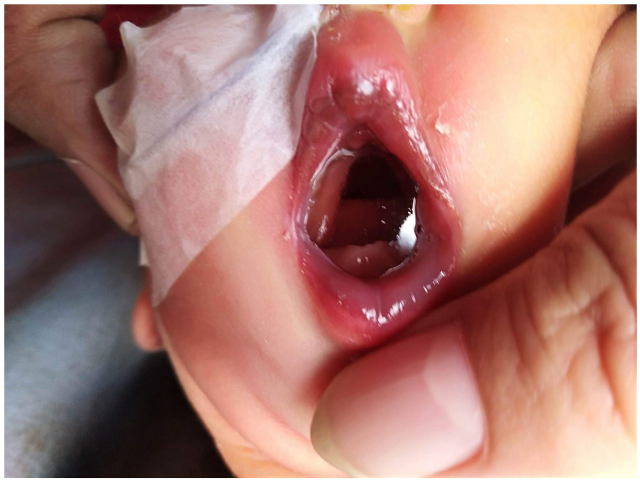

FIGURE 2 Pierre Robin syndrome in a child with no soft palate.

circulation and liver function that is not fully developed.[13] In our case, the neonate presented to the emergency with low birthweight, respiratory distress, and jaundice.

In the PRS, the risk of aspiration is higher because the newborn is not the same shape as other babies. Some examples are micrognathia, glossoptosis, a small oropharynx, mandibular hypoplasias, and a cleft palate. Our patient had an absence of soft palate which was rarely found in this PRS, with also micrognathy, and glossopitosis. PRS patients have two main problems: their airways get blocked, and they have trouble eating. Glossoptosis and the tongue moving back into the oropharynx block the upper airway in these patients, especially when they are eating. This leads to poor intake of food and may also lead to some case aspiration pneumonia.[14] Although they can be helpful in supplying nutrition, nasogastric tubes can also increase the risk of aspiration pneumonia. Upright feeding techniques, modifying the nipple for bottle feeding, using a nasogastric or orogastric feeding tube, and having a gastrostomy placed were all treatment options for feeding difficulties.[14] The most significant issue that can result in death in children with PRS is bronchial aspiration.[15] The respiratory issues of these patients must be managed. The treatment protocols consist of prone positioning with or without a nasopharyngeal tube, a laryngeal mask, prolonged intubation, tongue-lip adhesion, mandibular distraction osteogenesis, and tracheotomy.[14]

Local infections may be exacerbated by bronchitis and pneumonia. The infant becomes malnourished and undernourished and is frequently emaciated.[16] This condition is often accompanied by a palate abnormality or, which further compromises the patient's ability to take in adequate nutrition. Modern tongue-lip adhesion (TLA) and mandibular advancement (MA) surgery have completely changed the way PRS is treated. This aids in the child's nutrition and, if cleft palate or other palate abnormalities is present, leads to early correction of the condition.[17] Follow-up sessions with neonatologists (doctors who specialize in treating newborn babies) and the respiratory team are required for infants diagnosed with PRS. These appointments are necessary in order to check on the progress of any breathing or feeding issues. In infants diagnosed with PRS, repair of the palate abnormality is typically postponed until the child is approximately

TABLE 2 Treatment of the case.

Date	Hospital	Treatment	Status
30.01.2023	Rupgonj, Narayangonj College Hospital (NICU)	1. Oxygen Inhalation 2. Intravenous fluid 3. Inj. Ceftazidime 4. Inj. Amikacin 5. Inj. Dobutamine 6. Inj. Calcium gluconate	Patient take discharge on request (DOR)
04.02.2023	Maternal and Child Health Training Institute	1. Inj. Ampicillin (1.3 mL Intravenous—12 hourly) 2. Inj. Gentamicin (1.3 mL Intravenous—once daily) 3. 10% Baby saline (12 micro/drops)-Intravenous 4. Continuous phototherapy	Discharged

TABLE 3 Daily follow-up of the case in Maternal and Child Health Training Institute.

Day	Heart rate	Respiratory rate	Temperature	Heart	Lungs	Sp02	Jaundice
Day 6 7.00 PM	120/min irregular	50/min	98.1 F	Nothings abnormalities	Clear	95% without 2 L oxygen	Present
Day 7 10.00 AM	110/min irregular	48/min	98.2 F	Nothings abnormalities	Clear	99% with 2 L oxygen	Present
Day 8 3.30 PM	110/min irregular	40/min	98.0 F	Nothings abnormalities	Clear	95% with 2 L oxygen	Present
Day 9 8.00 PM	126/min irregular	50/min	98.0 F	Nothings abnormalities	Clear	95% with 2 L oxygen	Present
Day 10 4.00 PM	78/min regular	48/min	98.1 F	Nothings abnormalities	Mild creps	96% with 1 L oxygen	Present
Day 11 9.00 PM	79/min regular	46/min	98.2 F	Nothings abnormalities	Mild creps	97% with 1 L oxygen	Present
Day 12 9.00 AM	78/min regular	46/min	98.2 F	Nothings abnormalities	Clear	97% without oxygen	Present

18 months old and is scheduled after consultation with the entire cleft care team.

4 | CONCLUSION

Because of respiratory and nutrition difficulties, morbidity and mortality rates in PRS patients increased. The treatment of PRS patients necessitates diverse techniques. Yet, as a result of their anatomical defects, many patients do not live much longer.

AUTHOR CONTRIBUTIONS

Mohammad Ashraful Amin: Conceptualization; visualization; writing – original draft; writing – review and editing. **Taraque Ahamed Shawon:** Writing – original draft; writing – review and editing. **Naushad Khan Shaon:** Writing – review and editing. **Sabrina Nahin:** Visualization; writing – original draft; writing – review and editing. **Jannatul Fardous:** Writing – review and editing. **Mohammad Delwer Hossain Hawlader:** Supervision; writing – review and editing.

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

DATA AVAILABILITY STATEMENT

Data can be shared based on the reader's reasonable request and priority base, and some restrictions will apply.

ETHICS STATEMENT

The article is about a case study. As a result, our Ethics Committee's consent was not required.

CONSENT

The patient's parents had written informed consent taken for publishing this case report because of patient age's and images also were acquired.

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