

## CASE REPORT

# Challenges in Twin Pregnancy with Pierre Robin Sequence, Cardiomyopathy, and Post-Operative Respiratory Complications: A Case Report

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**ABSTRACT**

This case report discusses the successful management of a pregnant patient with a twin pregnancy, Pierre Robin Sequence (PRS), and peripartum cardiomyopathy (PPCM) who underwent a cesarean section. PRS, characterized by micrognathia, glossoptosis, and airway obstruction, presents significant challenges in anesthesia management, particularly concerning difficult intubation and airway management. PPCM, a condition marked by left ventricular systolic dysfunction and reduced ejection fraction, further complicates anesthesia due to the risk of myocardial depression. The patient's condition was exacerbated by pre-eclampsia and anemia. After a thorough preoperative evaluation, regional anesthesia was selected as the safest option, aiming to minimize hemodynamic fluctuations and reduce the risks of respiratory complications and opioid sensitivity. The use of inotropic support was crucial in maintaining cardiovascular stability throughout the perioperative period. This case highlights the importance of a multidisciplinary approach, involving obstetrics, anesthesiology, and cardiology, in the management of complex cases involving rare conditions. Regional anesthesia proved to be an effective strategy for minimizing risks and ensuring a successful outcome, emphasizing the need for

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individualized care in high-risk obstetric patients.

## KEYWORDS

• Pierre Robin Sequence • Peripartum Cardiomyopathy • Regional Anesthesia • Twin Pregnancy • Anesthesia Management

## INTRODUCTION

Pierre Robin Sequence (PRS) was first described by Dr. Pierre Robin, a French dental surgeon, in the early 20th century. PRS presents as a triad of micrognathia (small lower jaw), glossoptosis (displacement of the tongue to the back of the oral cavity), and airway obstruction. These features can occur either as an isolated condition or as part of a genetic syndrome. In approximately 60% of cases, PRS is associated with genetic syndromes such as Stickler, Velocardiofacial, and Treacher-Collins syndromes, necessitating preoperative echocardiography to assess potential cardiac involvement<sup>1</sup>. The pathophysiology of PRS begins with micrognathia, which leads to the displacement of the tongue into the nasopharynx, preventing the fusion of the palatal shelves and causing a cleft palate. This sequence of events results in airway obstruction and obstructive sleep apnea (OSA)<sup>2</sup>. The craniofacial abnormalities caused by PRS are typically detected at birth, where neonates present with respiratory distress (e.g., stridor, retractions, cyanosis), feeding difficulties, and failure to thrive. In addition to the craniofacial issues, PRS can be associated with cardiovascular abnormalities such as patent ductus arteriosus (PDA), right heart failure, pulmonary hypertension, and pulmonary stenosis, as well as neuromuscular and skeletal anomalies<sup>3</sup>.

The management of PRS during anesthesia is complex due to craniofacial abnormalities that make both ventilation and intubation difficult. In many cases, facemask ventilation can also be challenging. These patients are at increased risk of spontaneous airway collapse in the postoperative period. Furthermore, they exhibit increased sensitivity to opioids, necessitating careful dose adjustments. Feeding difficulties and gastroesophageal reflux contribute to a higher risk of aspiration, which requires precautionary measures

during anesthesia. A thorough preoperative assessment is essential for planning the anesthesia approach. This includes a detailed history and physical examination, focusing on airway evaluation and cardiopulmonary assessment. The airway examination should assess factors such as mouth opening, the presence of cleft palate, degree of airway obstruction, severity of micrognathia, and the presence of other craniofacial or skeletal abnormalities. Identifying features of OSA is crucial to predicting both intraoperative and postoperative airway complications<sup>4</sup>.

In addition to the complications associated with PRS, peripartum cardiomyopathy (PPCM) may present an additional challenge in the anesthetic management of pregnant patients. PPCM is a form of dilated cardiomyopathy that occurs late in pregnancy or early postpartum, characterized by left ventricular systolic dysfunction and a reduced ejection fraction. This condition can precipitate heart failure, especially during the perioperative period, making the management of anesthesia critical. The anesthesia approach for patients with PPCM is influenced by factors such as the urgency of surgery, the patient's hemodynamic status, and their New York Heart Association (NYHA) class. The primary goals of anesthesia management include avoiding myocardial depression, maintaining adequate preload, preventing increases in afterload, controlling tachycardia, and ensuring gradual titration of anesthetic agents to avoid sudden hypotension.<sup>5-7</sup>

Pregnant patients, especially those with PRS and PPCM, are at very high risk during surgery due to the physiological changes of pregnancy, which include increased cardiac output, a difficult airway, and a full stomach. The combination of these factors requires careful planning and management to avoid complications. General anesthesia can be

particularly challenging due to the difficult airway, the side effects of anesthetic drugs, and post-operative ventilation concerns. Furthermore, general anesthesia can lead to greater hemodynamic fluctuations, which can exacerbate the condition of a patient with PPCM. Therefore, neuraxial anesthesia is generally preferred, as it helps reduce afterload and improve cardiac output. However, it must be administered carefully, following ASRA guidelines, especially if the patient is on anticoagulants. Neuraxial anesthesia also helps reduce preload, alleviating hydrostatic pressure in the pulmonary capillaries and preventing the worsening of pulmonary edema.

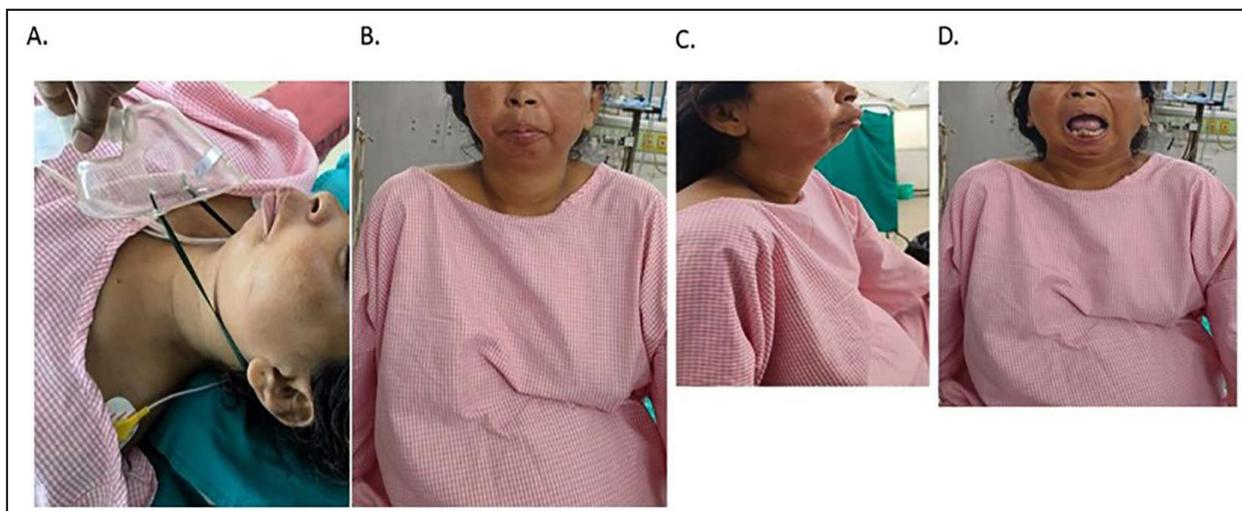
In cases of significant reduction in systemic vascular resistance (SVR), management should focus on the careful administration of fluids and vasopressors to maintain adequate coronary and organ perfusion. Graded neuraxial blocks can be safely used in cases where the ejection fraction is between 35% and 45%. This approach, when done cautiously, can provide the necessary anesthesia for

cesarean delivery while minimizing the risk of exacerbating cardiac dysfunction<sup>8</sup>.

The management of a pregnant patient with both Pierre Robin Sequence and peripartum cardiomyopathy requires a multifaceted approach. Thorough preoperative evaluation, careful anesthesia planning, and vigilant intraoperative and postoperative monitoring are critical to achieving a successful outcome.

### CASE REPORT

A 25-year-old female, gravida 2 para 1, weighing 50 kg and with a height of 155 cm, presented to the emergency operating theatre (O.T.) at 31 weeks gestation with a twin pregnancy (monochorionic monoamniotic). She was diagnosed with pre-eclampsia, moderate anemia, and Pierre Robin Sequence (PRS). The patient was admitted with complaints of difficulty breathing and a dry cough. Her relative reported a history of snoring. The patient's pregnancy had been uncomplicated until the third trimester, with no nausea, vomiting, or drug intake in the first trimester, and no history of headache, blurring of vision, or vaginal bleeding (Figure 1).



**Figure 1:** Showing difficult intubation (A) Case of Pierre robin sequence, (B) Micrognathia, (C & D) Reduced month opening, short neck

### Clinical Examination

Upon general examination, the patient exhibited pallor, pitting edema across the body, and face. She was well-oriented to time, place, and person. Respiratory examination revealed bilateral basal crepitations, while cardiovascular examination showed normal heart sounds (S1, S2) without murmurs. The

airway examination revealed a mouth opening of 2 fingers, Mallampati grade 4, glossoptosis, and micrognathia. The neck movement was restricted, and the thyromental distance was less than 6 cm. The spine was of normal shape, and all teeth were intact, though malaligned in the lower jaw.

### Vital Signs and Investigations

On presentation, the patient had a heart rate of 112/min, a blood pressure of 134/82 mmHg, a respiratory rate of 28/min, and an oxygen saturation (SpO<sub>2</sub>) of 97% on room air. The electrocardiogram (ECG) revealed left axis deviation and sinus tachycardia. A 2D echocardiogram (ECHO) showed aortic valve maximum velocity of 1.0 m/sec, normal chamber size, normal valves, moderate mitral regurgitation, and moderate tricuspid regurgitation (v/p = 3.5/50). There was global left ventricular hypokinesia, a left ventricular ejection fraction of 36%, and minimal pericardial effusion.

Blood investigations revealed hemoglobin (Hb) of 10 gm%, total leukocyte count (TLC) of 9800 cells/mm<sup>3</sup>, and platelet count of 1.63 lac/mm<sup>3</sup>. Liver and kidney function tests were normal.

### Preoperative Preparation

After obtaining high-risk consent, the patient was taken to the operating room, and standard monitoring was initiated. Baseline vital signs showed a heart rate of 110/min, blood pressure of 140/88 mmHg, respiratory rate of 30/min, and SpO<sub>2</sub> of 96% on room air. The patient's temperature was 37.5°C, and a Foley catheter was placed with 50 mL of urine output. Two wide bore 18 gauge IV lines were secured.

The patient was dyspneic at rest (NYHA Grade 4) and unable to lie flat on the operating table. Premedication included 4 mg of intravenous ondansetron and 1 gm of intravenous tranexamic acid in 50 mL 0.9% NaCl crystalloid solution. Oxygen was administered via a Hudson mask at 6 L/min. Inotropic support was initiated with dobutamine (1 mcg/ml) at 10 mcg/hour and noradrenaline (1 mcg/ml) at 2 mL/hour, adjusted to maintain the mean arterial pressure within ±20% of baseline.

### Anesthesia and Surgical Management

Regional anesthesia was deemed the most suitable option given the patient's high risk for cardio-respiratory complications and difficult airway. After explaining the procedure to the patient, spinal anesthesia was performed under sterile conditions. The patient was positioned sitting with knees resting on the edge of the bed and feet supported by a

stool. The skin was infiltrated with 3 mL of 2% lignocaine, and spinal anesthesia was administered at the L3-L4 intervertebral space using a 23-gauge Quincke needle. After free-flowing cerebrospinal fluid (CSF), 1.8 mL of 0.5% heavy bupivacaine and 0.2 mL of fentanyl were injected into the subarachnoid space. The patient was then immediately repositioned supine.

I.V. fluids (0.9% saline) were initiated at a rate of 50 mL/hour, and motor blockade was assessed using the modified Bromage scale. When the motor blockade score reached 4, the patient was handed over to the surgical team. A Pfannenstiel incision was made, and the twins were delivered within 10 minutes of the incision. Intraoperative hemodynamics showed a heart rate of 120/min, blood pressure of 136/86 mmHg, and SpO<sub>2</sub> of 100% on 6L/min oxygen.

Oxytocin was not administered, and after cord clamping, the placenta was delivered in a controlled manner. Hemostasis was achieved, and the uterus was well-contracted, as confirmed by the obstetrician. The estimated blood loss was approximately 500 mL, and urine output was 150 mL.

### Intraoperative Monitoring and Blood Gas Analysis

Intraoperative blood gas analysis showed the following results: pH 7.42, pCO<sub>2</sub> 25.8 mmHg, pO<sub>2</sub> 131 mmHg, SpO<sub>2</sub> 99.4%, K<sup>+</sup> 4.6 mmol/L, Na<sup>+</sup> 135 mmol/L, HCO<sub>3</sub><sup>-</sup> 16.5 mmol/L, Hb 9.5 gm/dL, and lactate 1.4 mmol/L.

### Post-operative Period

After completing the procedure, the patient was transferred to the postoperative anesthesia recovery room. Inotropic support (noradrenaline and dobutamine) was gradually tapered. Postoperative vital signs showed a heart rate of 115/min, blood pressure of 130/80 mmHg, and SpO<sub>2</sub> of 100% on 6L/min oxygen. Postoperative blood gas analysis revealed the following: pH 7.40, pCO<sub>2</sub> 31.5 mmHg, pO<sub>2</sub> 141 mmHg, SpO<sub>2</sub> 99.5%, K<sup>+</sup> 4.5 mmol/L, Na<sup>+</sup> 136 mmol/L, HCO<sub>3</sub><sup>-</sup> 20.5 mmol/L, Hb 9.0 gm/dL, and lactate 1.3 mmol/L.

This case highlights the complexity of managing a patient with multiple coexisting conditions, including Pierre Robin Sequence, pre-eclampsia, anemia, and peripartum cardiomyopathy, requiring careful anesthesia

and surgical planning. The decision to proceed with spinal anesthesia was based on the need to minimize hemodynamic fluctuations and reduce the risk of respiratory complications associated with general anesthesia. The use of inotropic support was crucial in

maintaining hemodynamic stability during the perioperative period. Postoperatively, close monitoring of the patient's cardiovascular and respiratory status was essential for a successful recovery.

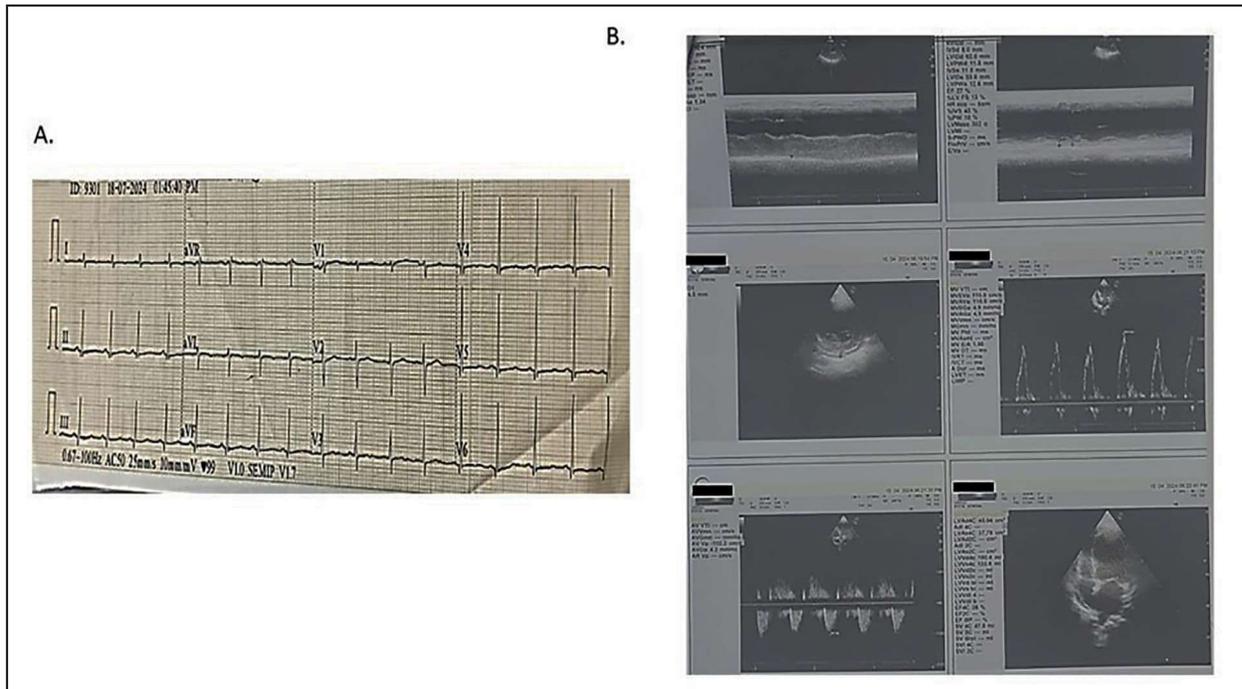


Figure 2: Diagnostic Figures: (A) ECG, (B) ECHO

## DISCUSSION

The incidence of Pierre Robin Sequence (PRS) ranges from 1 in 5,000 to 1 in 85,000 births. The severity of symptoms can vary widely, with some cases going unrecognized. In addition to clinical examination, radiological tests are often used to assess potential bony and soft tissue abnormalities associated with PRS<sup>9</sup>.

Airway management in PRS patients can be challenging for anesthetists. The normal maxillo-facial angle is less than 90°, but when it exceeds 100°, visualizing the glottis during direct laryngoscopy becomes difficult<sup>10</sup>. Airway obstruction and difficult intubation can lead to perioperative complications such as respiratory distress, hypoxia, and respiratory failure. PRS patients are particularly sensitive to opioids, as chronic airway obstruction and hypoxia increase opioid sensitivity<sup>4</sup>.

Similarly, Semjen F *et al.* demonstrated the use of a paraglossal approach combined with a gum elastic bougie for intubation in infants with PRS, highlighting the difficulty of

intubating these patients<sup>11</sup>. Satoru Takeshita *et al.* reported that fiberoptic nasotracheal intubation was required to manage severe dyspnea due to upper airway obstruction immediately after birth in PRS patients<sup>12</sup>. Philip Michele BA *et al.* presented an adult patient with PRS who continued to face airway difficulties, suggesting that airway management challenges persist with age<sup>13</sup>.

Severe airway obstruction is a significant risk factor for postoperative morbidity and mortality. Postoperative complications such as ventilator-induced lung injury and pulmonary infections can arise from prolonged mechanical ventilation following general anesthesia. Therefore, considering the risk-benefit ratio, regional anesthesia was selected as the preferred approach in our case.

In patients with dilated cardiomyopathy, pregnancy is generally not advised, or termination is recommended during the first trimester. In these patients, left and right ventricular systolic dysfunction leads

to progressive cardiac enlargement, a process known as remodeling, which causes symptoms of congestive heart failure (CHF). Pregnancy exacerbates these symptoms, particularly during the third trimester, due to increased intravascular volume and metabolic demand. Neuraxial anesthesia induced changes in preload and after load align with the therapeutic goals in such cases. Therefore, spinal anesthesia was chosen for our patient.

Studies by Shnaider *et al.*<sup>14</sup>, Kotekar *et al.*<sup>15</sup>, and Nallam *et al.*<sup>16</sup> demonstrated the successful use of spinal or epidural anesthesia in patients with a left ventricular ejection fraction (LVEF) as low as 20%, 41%, and 29%, respectively. In these patients, regional anesthesia improved myocardial performance by reducing after load on the left ventricle, although it did not enhance contractility.

## CONCLUSION

Pierre robin syndrome is a rare disease, which can have serious consequences. PPCM is again a rare entity complicating the terminal stages of pregnancy. Our patient had combination of all the above factors. Successful management in the form of regional anaesthesia was done in a case of patient with twin pregnancy with pierre robin sequence with cardiomyopathy with cardiac and respiratory complications posted for cesarean section. The successful management of this case demonstrates the importance of a multidisciplinary approach in the management of complex obstetric cases involving rare conditions like PRS and PPCM. Regional anesthesia provided a safe and effective alternative to general anesthesia, allowing for better control of hemodynamic parameters while managing the patient's difficult airway and underlying cardiac condition. Close collaboration among the obstetric, anesthesiology, and cardiology teams was essential to achieving a successful outcome, ensuring patient safety, and minimizing the risks of perioperative complications.

## REFERENCES

1. Mackay D.R. Controversies in the diagnosis and management of the Robin sequence. *J Craniofac Surg.* 2011 Mar; 22(2): 415-20. PubMed
2. Prows C.A., Bender P.L. Beyond Pierre Robin sequence. *Neonatal Netw.* 1999 Aug; 18(5):

- 13-9. PubMed
3. Hegde N., Singh A. Anesthetic Consideration in Pierre Robin Sequence. Updated 2023 Jul 31. In: StatPearls Internet. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK576442/>
4. Brown K.A., Laferrière A., Moss I.R. Recurrent hypoxemia in young children with obstructive sleep apnea is associated with reduced opioid requirement for analgesia. *Anesthesiology.* 2004 Apr; 100(4): 806-10; discussion 5A. PubMed Reference list
5. Monasterio F.O., Molina F., Berlanga F., López M.E., Ahumada H., Takenaga RH, Ysunza A. Swallowing disorders in Pierre Robin sequence: its correction by distraction. *J Craniofac Surg.* 2004 Nov; 15(6): 934-41. PubMed Reference list
6. Shobha V., Mittal S., Rawat K.V., Anaesthetic management in Pierre Robin syndrome with bicuspid aortic valve: A case report. *Indian J Clin Anaesth* 2019;6(3):473-475
7. Cladis, Franklyn, Kumar A., Lorelie G., Todd O., Mathew F. *et al.* Pierre Robin Sequence: A Perioperative Review. *Anesth Analgesia* 2014; 119(02): 400-12.
8. Breugem C., Courtemanche D. Robin Sequence: Clearing Nosologic Confusion. *Cleft Palate Craniofac J* 2010; 47: 197- 200.
9. Gangopadhyay N. Pierre Robin Sequence. *SeminPlastSurg* 2012; 26(2); 76-80.
10. Yosha Prasad: ATOTW 250 - The Difficult Paediatric Airway; [www.totw.anaesthesiologist.org](http://www.totw.anaesthesiologist.org), 13/02/2012: Page 1of 13
11. Semjen F., Bordes M., Cros A.M. Intubation of infants with Pierre Robin syndrome: the use of the paraglossal approach combined with a gum-elastic bougie in six consecutive cases. *Anaesthesia.* 2008; 63(2): 147-50.
12. Takeshita S., Ueda H., Goto T., Muto D., Kakita H., Oshima K., Tainaka T., Ono T., Kazaoka Y., Yamada Y. Case report of Pierre Robin sequence with severe upper airway obstruction who was rescued by fiberoptic nasotracheal intubation. *BMC Anesthesiol.* 2017 Mar 14; 17(1): 43. doi: 10.1186/s12871-017-0336-0. PMID: 28288578; PMCID: PMC5348775.
13. Philip M., Patel S., Chaudhry F., Napoli S., Pandya Shah S. Airway Management May Not Improve in Adult Patients With Pierre Robin Sequence: A Case Report. *A Pract.* 2024 Aug 13;18(8):e01838. doi: 10.1213/

- XAA.0000000000001838. PMID: 39137102.
14. Shnaider R., Efri T., Sjmuk P., Larson S., Warters R.D., Katz J. Combined spinal - epidural anaesthesia for cesarean section in a patient with peripartum dilated cardiomyopathy. *Can J Anaesth* 2001; 48; 681-3.
  15. Kotekar N., Nagalakshmi N.V., Chandrashekar. A rare case of peripartum cardiomyopathy posted for caesarean section. *Indian J Anaesth* 2007; 51: 60-4.
  16. Nallam S.R., Kosinapalle S., Jyothirmai I., Ratnamaiah A., Reddy K.S. Perioperative anaesthetic management of a case of severe dilated cardiomyopathy undergoing elective lower segment caesarean section under epidural anaesthesia. *Int J Sci Stud* 2014; 2: 225-7.