DOI: 10.21088/ijaa.2349.8471.13426.3

Indian Journal of Anesthesia and Analgesia
Volume 13 Number 4 October-December 2026

CASE REPORT

Syndromic Facies, Distorted Spine, and Anticipated Difficult Obstetric Airway, Anaesthetic Challenges in an Emergency Caesarean Section

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HOW TO CITE THIS ARTICLE:

Sukriti Atram, Vinayak Jadhav, Soniya et. al, Syndromic Facies, Distorted Spine, and Anticipated Difficult Obstetric Airway, Anaesthetic Challenges in an Emergency Caesarean Section. Ind J Anesth Analg. 2026; 13(4): 183-188.

ABSTRACT

Background: Undiagnosed syndromic conditions in obstetric patients can significantly complicate anaesthetic management, particularly during emergency situations. Anatomical variations, spinal deformities, and altered physiological responses may predispose to regional anaesthesia failure and airway difficulty.

Case Presentation: We report a case of a 31-year-old primigravida with unrecognized syndromic features who underwent emergency caesarean section. Routine preoperative investigations were within normal limits. Initial spinal anaesthesia failed, necessitating conversion to general anaesthesia. Mask ventilation was difficult due to facial dysmorphism and required a two-person technique. Anticipated difficult airway management with video laryngoscopic intubation was successfully performed. Both maternal and neonatal outcomes were favourable.

Conclusion: This case underlines the necessity of meticulous preoperative assessment, vigilance for syndromic features, and preparedness for airway challenges and anaesthetic conversion in obstetric emergencies.

KEYWORDS

• Difficult Airway • Syndromic Parturient • Emergency Caesarean Section • Video Laryngoscopy • Failed Spinal Anaesthesia • Obstetric Anaesthesia

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➢ **Received:** 03-08-2026 ➢ **Accepted:** 08-09-2026

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INTRODUCTION

Syndromic disorders often present with craniofacial and musculoskeletal abnormalities that complicate both airway management and the administration of regional anaesthesia.[1] In emergency obstetric settings, the absence of a prior diagnosis can further amplify these challenges.[2] Limited time for comprehensive evaluation necessitates rapid, evidence-based anaesthetic decision-making. Pregnancy itself induces physiological changes that reduce functional residual capacity, increase oxygen consumption, and heighten the risk of rapid desaturation during airway management.[3]

The incidence of difficult intubation in obstetric patients ranges from 1:250 to 1:750, which is significantly higher than in the general surgical population.[4] When compounded by undiagnosed syndromic features—such as craniofacial dysmorphism, spinal deformities, and restricted neck mobility—the anaesthetic complexity increases substantially. Furthermore, regional anaesthesia techniques may fail due to anatomical distortion, necessitating rapid conversion to general anaesthesia with attendant airway risks.[5]

We present a rare case of an undiagnosed syndromic parturient encountered during emergency caesarean section, illustrating the perioperative challenges and key lessons for safe anaesthetic care in such high-risk scenarios.

CASE PRESENTATION

A 31-year-old primigravida at 38 weeks' gestation with an uneventful antenatal course was posted for emergency lower segment caesarean section due to fetal distress with non-reassuring cardiotocography. She had no documented comorbidities, no history of previous surgery or anaesthesia, and had received regular antenatal care without identified complications.

Preoperative Assessment

On pre-anaesthetic evaluation, the patient was alert, cooperative, and hemodynamically stable, with a blood pressure of 128/82 mmHg, a heart rate of 92 beats per minute, and an oxygen saturation of 98% on room air. Routine preoperative investigations, including complete blood count (hemoglobin 11.2 g/dL), liver function tests, renal function tests, random blood glucose, and a coagulation profile, were all within normal limits.

Physical examination revealed striking syndromic features: short stature (height approximately 142 cm), a markedly kyphotic thoracic spine, hypoplastic maxilla, prominent prognathic mandible, macroglossia, and a short, thick neck with severely restricted extension. Airway assessment demonstrated a Mallampati grade IV view, a thyromental distance of less than 6 cm, limited mouth opening (approximately 3 cm), and poor neck mobility. A positive family history was elicited the patient's mother exhibited similar facial and skeletal features, suggesting a hereditary syndromic condition, although no formal genetic diagnosis had been established in either individual.



Figure 1: Spine deformity with rib hump and asymmetry



Figure 2: craniofacial abnormalities (hypoplastic maxilla, asymmetry of the eye and upper face, flattened mid face, abnormal jaw protution prominent prognathic mandible, micrognathia, restricted mouth opening)

Anaesthetic Management

Given the spinal deformity and the urgency of the obstetric indication, a decision was made to attempt spinal anaesthesia initially. Informed consent was obtained after explaining the risks, benefits, and the possibility of conversion to general anaesthesia. The patient was positioned in the sitting position with maximal flexion. Using strict aseptic precautions, spinal anaesthesia was attempted at the L3–L4 interspace using a 25-gauge Quincke spinal needle. Due to the severe kyphosis and anatomical distortion, multiple attempts were required. Although cerebrospinal fluid was obtained and 2.2 mL of 0.5% hyperbaric bupivacaine with 25 mcg fentanyl was administered, the resultant block was inadequate, achieving only a T10 sensory level bilaterally with incomplete motor blockade after 15 minutes.



Figure 3: short stature, Syndactyly fingers and toes



Figure 4: Two-person mask technique

Given the inadequate spinal block, hemodynamic stability of the parturient, and the urgency of fetal delivery, the decision was made to proceed with general anaesthesia. The difficult airway trolley was prepared with a video laryngoscope, bougie, supraglottic airway devices, an emergency cricothyrotomy kit, and a fiberoptic bronchoscope on standby.

Standard American Society of Anesthesiologists monitoring was applied. Preoxygenation was initiated with 100% oxygen via a face mask. However, achieving an effective mask seal proved extremely difficult due to the patient's hypoplastic maxilla, prognathism, and macroglossia. Initial single-operator mask ventilation resulted in significant air leak and inadequate chest rise. Two anaesthesiologists were then required to hold the mask with both hands, applying maximal pressure to achieve an effective seal, while a third provider performed bag ventilation. Only with this three-person technique and firm mask-hold was adequate mask ventilation achieved. Rapid sequence induction was performed with intravenous propofol 2 mg/kg and succinylcholine 1.5 mg/kg following pre-treatment with glycopyrrolate 0.2 mg. Cricoid pressure was applied. Direct laryngoscopy was not attempted given the anticipated difficult airway. Video laryngoscopy using the C-MAC® system was performed, revealing a Cormack-Lehane grade III view. A gum elastic bougie was introduced under direct video visualization and railroaded through the glottis. A 7.0 mm internal diameter cuffed endotracheal tube was then advanced over the bougie into the trachea. Correct tube placement was confirmed by capnography showing an end-tidal CO_2 waveform, bilateral chest auscultation, and observation of chest rise.

Anaesthesia was maintained with oxygen, air (FiO_2 0.5), and sevoflurane (1.0–1.2 minimum alveolar concentration). Continuous monitoring of maternal vital signs, uterine tone, and estimated blood loss was performed throughout the procedure.

Surgical Outcome

The surgical procedure was completed uneventfully over 45 minutes. A healthy male neonate weighing 2.8 kg was delivered with Apgar scores of 8 at one minute and 9 at five minutes. The neonate required no resuscitative measures and was handed over to the attending pediatrician in stable condition. Estimated blood loss was approximately 450 mL, and uterine tone was satisfactory following administration of an oxytocin infusion.

Postoperative Course

At the conclusion of surgery, neuromuscular blockade was reversed with neostigmine 2.5 mg and glycopyrrolate 0.5 mg intravenously. The patient was extubated after confirming full return of neuromuscular function, adequate spontaneous respiratory effort, protective airway reflexes, and hemodynamic stability. Extubation was smooth without complications.

The patient was transferred to the post-anaesthesia care unit for monitoring. Postoperative analgesia was provided with intravenous paracetamol 1 g every 6 hours and diclofenac 75 mg intramuscularly every 12 hours. Pain was well controlled, and the patient mobilized on postoperative day one. Recovery was uneventful, and she was discharged on postoperative day three in good condition with the neonate.

A detailed discussion was held with the patient and her family regarding the syndromic features observed. Referral to a clinical geneticist was made for comprehensive evaluation and definitive diagnosis, along with counseling regarding future pregnancies and anaesthetic implications.

DISCUSSION

This case highlights several critical aspects of anaesthetic management in undiagnosed syndromic parturients presenting for emergency obstetric intervention. The combination of craniofacial abnormalities, spinal deformity, and the physiological changes of pregnancy created a scenario of compounded anaesthetic risk requiring careful planning, anticipation of complications, and readiness for rapid intervention.

Difficult Airway in Syndromic Patients

Syndromic conditions associated with craniofacial dysmorphism frequently result in difficult mask ventilation and intubation.[1] Features such as hypoplastic maxilla, mandibular prognathism, macroglossia, and limited neck extension are well-established predictors of airway difficulty.[6] In our patient, conventional single-operator mask ventilation was ineffective due to severe facial dysmorphism and macroglossia, leading to significant air leak and poor chest expansion. This necessitated escalation to a three-person approach in which two anaesthesiologists applied a tight two-handed mask seal with maximal pressure while a third provider delivered bag ventilation. Such escalation from one-person to multi-person mask ventilation is consistent with difficult airway guidelines that emphasise optimising facemask seal, using additional personnel, and employing airway adjuncts to maintain oxygenation in obstetric patients at high risk of rapid desaturation.[7]

The use of video laryngoscopy has revolutionized difficult airway management, particularly in obstetric anaesthesia where time constraints and physiological derangements limit the margin for error.[8] Studies have demonstrated that video laryngoscopy improves glottic visualization, reduces airway trauma, and increases first-attempt intubation success rates compared to direct laryngoscopy in difficult airway scenarios.[9] In our case, the C-MAC® video laryngoscope combined with bougie-guided intubation allowed successful first-attempt intubation despite a Cormack-Lehane grade III view, thereby minimizing maternal hypoxemia and fetal compromise.

Failed Spinal Anaesthesia in Spinal Deformity

Regional anaesthesia is the preferred technique for caesarean section due to reduced maternal aspiration risk, avoidance of neonatal exposure to general anaesthetic agents, and lower overall maternal morbidity.[10] However, in patients with spinal deformities such as kyphosis, scoliosis, or syndromic spinal abnormalities, the technical success rate of neuraxial blockade is significantly reduced.[5] Anatomical distortion can lead to difficulty in identifying landmarks, multiple attempts, inadequate spread of local anaesthetic, and patchy or incomplete blocks.[11]

In our patient, despite successful dural puncture and administration of an appropriate dose of hyperbaric bupivacaine, the resulting sensory block was inadequate. This may be attributed to altered cerebrospinal fluid dynamics, abnormal spinal canal anatomy, or unpredictable local anaesthetic spread in the presence of kyphotic deformity.[12] The decision to convert to general anaesthesia was made promptly to avoid maternal discomfort, psychological distress, and potential surgical complications from inadequate anaesthesia.

Obstetric Airway: Unique Challenges

Pregnancy induces a constellation of physiological changes that complicate airway management. Increased progesterone

levels cause mucosal oedema and capillary engorgement of the upper airway, reducing airway diameter and increasing friability.[13] The gravid uterus elevates the diaphragm, reducing functional residual capacity by approximately 20%, which accelerates oxygen desaturation during apnoea.[14] Additionally, increased oxygen consumption (up to 20% above baseline) further shortens the safe apnoea time.[15]

These physiological alterations are compounded in syndromic patients with pre-existing anatomical airway compromise. The combination demands meticulous preoxygenation, avoidance of prolonged laryngoscopy attempts, and immediate availability of rescue airway devices. In our case, effective preoxygenation using a two-person mask technique and prompt use of video laryngoscopy ensured maternal oxygenation was maintained throughout the airway management process.

Recognition of Undiagnosed Syndromes

Several genetic syndromes associated with craniofacial and skeletal abnormalities may remain undiagnosed into adulthood, particularly in resource-limited settings or in the absence of significant functional impairment during childhood.[16] Syndromes such as Klippel-Feil (characterized by congenital fusion of cervical vertebrae, short neck, and restricted neck mobility), Pierre Robin sequence (micrognathia, glossoptosis, and cleft palate), Treacher Collins syndrome (mandibulofacial dysostosis), Goldenhar syndrome (hemifacial microsomia), and Marfan-like phenotypes may present for the first time during pregnancy when anaesthetic intervention is required.[17]

While our patient did not receive a definitive genetic diagnosis, the constellation of findings, such as short stature, kyphotic spine, hypoplastic maxilla, prognathism, macroglossia, and a short neck, along with a positive family history, suggests a heritable syndromic condition. Recognition of such features during the pre-anaesthetic assessment is crucial for anticipating potential complications and formulating an appropriate management plan.

Multidisciplinary Approach and Preparedness

Safe anaesthetic management in complex obstetric cases requires effective communication and collaboration among obstetricians, anaesthesiologists, neonatologists, and nursing staff.[18] In this case, early recognition of potential airway difficulty prompted preparation of the difficult airway trolley, availability of experienced personnel, and readiness for immediate neonatal resuscitation.

Institutional preparedness—including availability of advanced airway equipment, training in video laryngoscopy and fibreoptic techniques, and regular simulation-based difficult airway drills—is essential to manage rare but high-risk scenarios.[19] The presence of a written difficult airway algorithm and adherence to established guidelines (such as those from the Difficult Airway Society or the American Society of Anesthesiologists) can guide decision-making and improve outcomes.[20]

Implications for Future Pregnancies

Following this case, genetic counselling was strongly recommended. Establishing a definitive diagnosis has implications not only for the patient's future anaesthetic care but also for genetic counselling regarding recurrence risk in future offspring and planning for subsequent deliveries. Ideally, such patients should be evaluated in a high-risk obstetric anaesthesia clinic during the antenatal period, allowing for thorough assessment, airway imaging if indicated, and formulation of a documented anaesthetic plan to guide management during labour or elective delivery.[21]

CONCLUSION

Undiagnosed syndromic parturients present formidable challenges to anaesthetic management, particularly in emergency obstetric situations where time is limited and physiological reserves are compromised. This case demonstrates the critical importance of thorough pre-anaesthetic evaluation, early recognition of dysmorphic features suggestive of syndromic conditions, and anticipation of both failed regional anaesthesia and difficult airway management.

Key learning points include the necessity of a low threshold for general anaesthesia in patients with spinal deformities and difficult airway predictors, the value of video laryngoscopy and advanced airway adjuncts in managing anticipated difficult intubation, and the importance of a two-person mask ventilation technique when facial

anatomy precludes an effective mask seal. Multidisciplinary coordination, institutional preparedness, and adherence to difficult airway algorithms are essential to ensure optimal maternal and neonatal outcomes.

Awareness of subtle syndromic features and a proactive, systematic approach to airway management can significantly reduce perioperative morbidity and mortality in these complex, high-risk obstetric cases.

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