

Case Report

Volume 13 Issue 5 - February 2025
DOI: 10.19080/JAICM.2025.13.555871

J Anest & Inten Care Med

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Anesthetic Management of a Pregnant Woman with VACTERL Syndrome and Vaginal Agenesis Undergoing Cesarean Section: A Case Report



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Submission: January 28, 2025; **Published:** February 04, 2025

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Keywords: VACTERL Syndrome; Cesarean Section; Anesthetic Management; Obstetric Anesthesia

Abbreviations: IH: Intracranial Hypertension; MRI: Magnetic Resonance Imaging; CSE: Combined Spinal-Epidural

Introduction

VACTERL syndrome is a rare association of congenital malformations involving at least three of the following anomalies: vertebral defects, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb defects. The estimated incidence ranges from 1 in 10,000 to 1 in 40,000 live births, with no identified genetic or environmental causes. Patients with VACTERL syndrome often pose significant challenges for anesthetic management due to their complex anatomy, comorbidities, and high risk of perioperative complications [1,2].

These patients often require multidisciplinary care throughout their lives due to the multisystemic nature of the syndrome. Vertebral anomalies, such as scoliosis, may complicate the administration of neuraxial anesthesia and pose challenges for airway management. Renal malformations can lead to fluid and electrolyte imbalances, while cardiac and tracheoesophageal anomalies increase the risk of intraoperative hemodynamic instability and aspiration. Proper anesthetic planning must take these considerations into account to reduce perioperative morbidity and mortality [3].

Case Description

We present the case of a 38-year-old female patient with

multiple congenital malformations consistent with VACTERL syndrome. The patient underwent a scheduled cesarean section at 38 weeks of gestation due to the inability to deliver vaginally as a result of a surgically reconstructed neovagina. The patient's medical history included severe lumbar scoliosis with restrictive pulmonary function (total lung capacity at 68%), unilateral renal agenesis, unicornuate uterus, and corrected vaginal atresia, with no associated cardiac anomalies. Anesthetic management for cesarean delivery in patients with VACTERL syndrome is complex and not well-standardized [3,4], making this case of particular interest.

Regarding the clinical course during her pregnancy, our patient was closely monitored by a specialized high-risk obstetric unit at Hospital Universitario La Paz (Madrid, Spain). Additionally, the patient experienced persistent headaches during the second trimester, prompting a neurological evaluation at 18 weeks of gestation. Differential diagnosis included intracranial hypertension (IH) due to possible cervical vertebral anomalies associated with VACTERL syndrome [5]. Magnetic resonance imaging (MRI) ruled out IH and confirmed hypnic headache, effectively managed with acetaminophen and caffeine. The patient's significant history of lumbar scoliosis required meticulous anesthetic planning for performing a neuraxial technique (Figure 1).



Figure 1: Posteroanterior chest X-ray performed prior to pregnancy showing thoracolumbar scoliosis with right dorsal convexity and pectus carinatum.

Obstetric and Anesthetic Management

At 34 weeks, fetal growth and Doppler studies were normal, and a cesarean section was scheduled for 38 weeks. The anesthetic approach included combined spinal-epidural (CSE) anesthesia using a Spinocan® 27G spinal needle and a Pencan® 18G epidural needle. Intrathecal administration of 7 mg of bupivacaine and 20 mcg of fentanyl achieved a sensory block up to T4. The epidural catheter allowed reinforcement with 5 ml of 2% lidocaine due to slight discomfort during uterine closure and facilitated postoperative analgesic management.

The procedure was uneventful, maintaining hemodynamic stability with a low-dose phenylephrine infusion (0.3-0.4mcg/kg/min) and an intraoperative blood loss of 500 ml. Postoperative pain management included epidural boluses of 0.2% ropivacaine and 2mg of morphine chloride. The team paid special attention to positioning during surgery to mitigate the risk of respiratory compromise due to scoliosis. Fluid management was carefully adjusted to prevent fluid overload, given the patient's single functional kidney. These measures contributed to a successful anesthetic outcome (Figure 2).

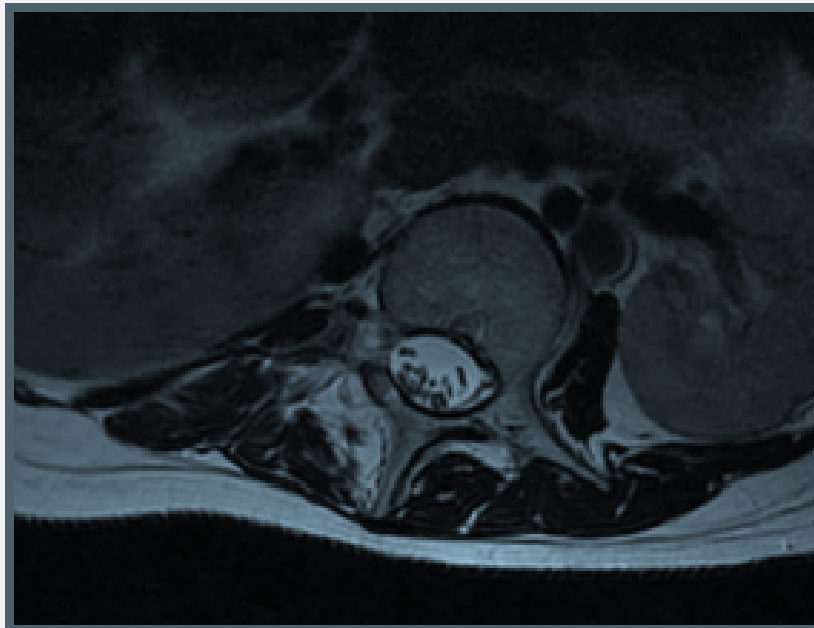


Figure 2: Lumbar spine MRI in T2 sequence demonstrating marked lumbar levoscoliosis. The same study reveals possible situs inversus (spleen on the right), right kidney agenesis, and left kidney ptosis, consistent with VACTERL syndrome.

Discussion

Patients with VACTERL syndrome pose unique challenges for anesthesiologists. Vertebral anomalies and scoliosis can complicate neuraxial anesthesia placement and lead to unpredictable drug distribution. Renal anomalies may affect fluid management and drug clearance. Additionally, tracheoesophageal and cardiac defects, though absent in this patient, often necessitate careful airway and hemodynamic management. Multidisciplinary collaboration is essential to optimize perioperative care and address these challenges [1,4,5]. The use of a combined spinal-epidural technique provided adequate, tailored surgical

anesthesia while minimizing the risks associated with general anesthesia, particularly in patients with potential respiratory compromise due to scoliosis. The epidural catheter facilitated effective postoperative analgesia, contributing to an optimized, pain-free recovery. Multidisciplinary planning was crucial in this case. Input from obstetricians, anesthesiologists, and neurologists ensured that the patient's unique anatomical and physiological challenges were addressed. Preoperative imaging studies, including MRI, helped exclude intracranial pathology and guided safe anesthetic management. The absence of cardiac anomalies simplified intraoperative monitoring and management but did not lessen the need for vigilance (Figure 3).

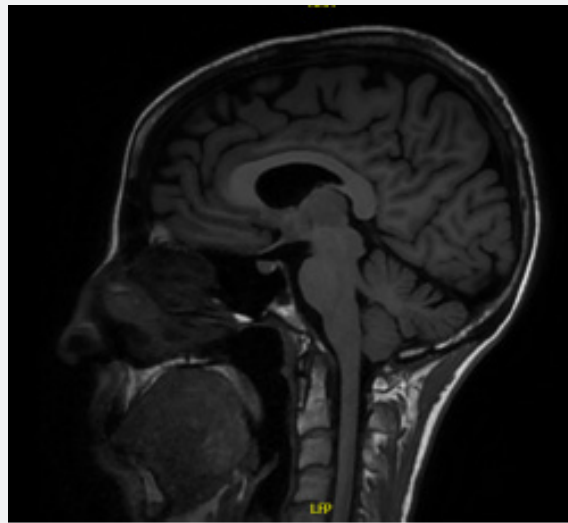


Figure 3: Cranial MRI in T1 sequence performed at 20 weeks of gestation showing permeable ventricular system and venous sinuses, mild right tonsillar ectopia, minimal ventricular asymmetry, slightly enlarged left Meckel's cave, and inversion of cervical lordosis, possibly postural, without significant abnormalities or evidence of intracranial hypertension.

Postoperative Course

The patient recovered without complications, including the absence of post-dural puncture headache or urinary retention. Effective postoperative analgesia provided by the epidural catheter contributed to early mobilization and discharge. Regular follow-up appointments were scheduled to monitor renal function and assess any long-term sequelae of the patient's condition.

Conclusion

This case underscores the importance of thorough planning in the management of obstetric patients with VACTERL syndrome. The absence of cardiac anomalies and effective multidisciplinary collaboration facilitated optimal perioperative care. An individualized approach, including tailored anesthetic techniques and meticulous perioperative management, is essential for improving outcomes in these high-risk patients.

Ethical Standards Compliance

Written informed consent was obtained from the patient for the publication of this case.

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DOI: [10.19080/JAICM.2025.13.555871](https://doi.org/10.19080/JAICM.2025.13.555871)

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