PAR Perioperative Anesthesia Reports

CASE REPORT

Anesthetic management of a patient with Fahr's disease, dextrocardia, and right lung agenesis: a case report

Lucas Lessa Nunes[1], Tiago Silva e Silva[1], José Fernando Amaral Meletti[1]

How to cite: Nunes LL, Silva TS, Meletti JFA. Anesthetic management of a patient with Fahr's disease, dextrocardia, and right lung agenesis: a case report. Periop. Anesth. Rep. 2026;4:e00032025. https://doi.org/10.61724/2965-3681/e00032025

ABSTRACT

Fahr's disease is a rare neurological disorder characterized by bilateral brain and vascular calcifications, often presenting with parkinsonism, cognitive decline, and neuromuscular abnormalities. We report the case of a 42-year-old male with Fahr's disease, right lung agenesis, and dextrocardia undergoing elective laparoscopic cholecystectomy. General anesthesia was induced with fentanyl, lidocaine, propofol, and rocuronium, maintaining intraoperative stability. Despite the challenges posed by pneumoperitoneum and pre-existing conditions, the patient was extubated without complications. This report demonstrates that standard anesthetic practices and intraoperative monitoring ensured a safe outcome in a patient with Fahr's disease, dextrocardia, and right lung agenesis, highlighting the adequacy of conventional strategies even in complex clinical scenarios.

KEYWORDS

Anesthesia; dextrocardia; hypocalcemia; vascular calcification

INTRODUCTION

Fahr's disease, also known as primary familial brain calcification, is a rare neurodegenerative disorder (<1:1.000.000)[1] characterized by abnormal calcium deposits in the basal ganglia, cerebellum, and subcortical white matter. It presents with a spectrum of neurological symptoms, including tremors, dystonia, cognitive decline, and psychiatric disturbances. Despite its long-recognized pathology, there is a scarcity of literature on its anesthetic implications. Dextrocardia and pulmonary agenesis are also rare congenital anomalies and may significantly impact perioperative monitoring and ventilation. Dextrocardia, in particular, complicates electrocardiographic interpretation, and pulmonary agenesis poses challenges to ventilation under pneumoperitoneum conditions. This case report describes the perioperative challenges and anesthetic management of a patient with Fahr's disease, dextrocardia, and right lung agenesis, aiming to highlight key considerations to optimize patient safety and outcomes. Written informed consent was obtained from the patient. The authors declare no conflict of interest.

CASE REPORT

A 42-year-old male patient, weighing 70 kg, with a medical history of Fahr's disease, dextrocardia, and right lung agenesis, was scheduled for elective laparoscopic cholecystectomy (Figures 1 and 2). This case describes the anesthetic management and perioperative challenges presented by this unique combination of conditions. His clinical manifestations of Fahr's disease included hand tremors and mild cognitive decline. Additionally,

[1] Faculdade de Medicina de Jundiaí, Centro de Ensino e Treinamento da Disciplina de Anestesiologia, Jundiaí, SP, Brasil

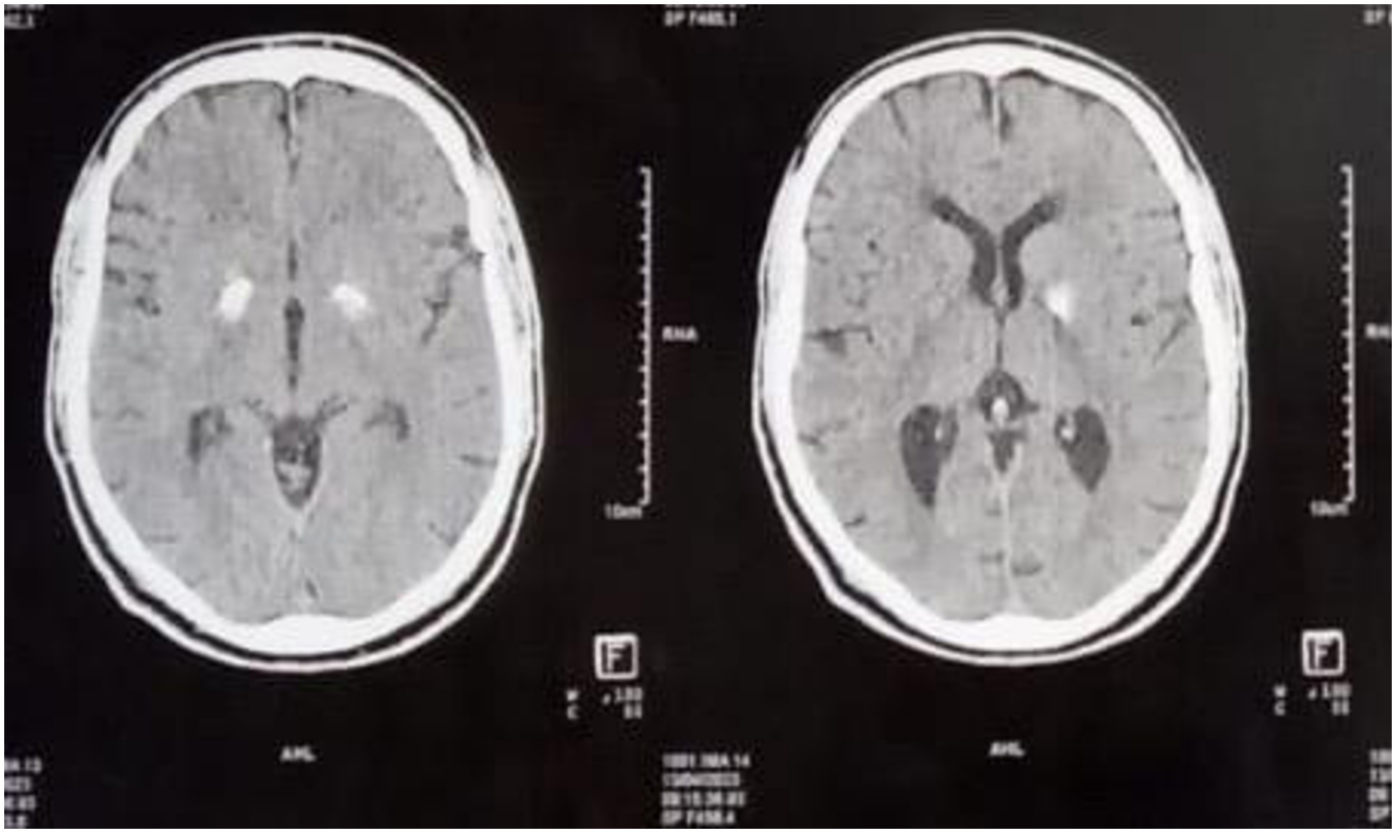

Figure 1. Axial CT scan of the patient's skull showing hyperattenuating images in the basal nuclei bilaterally, suggesting calcifications.

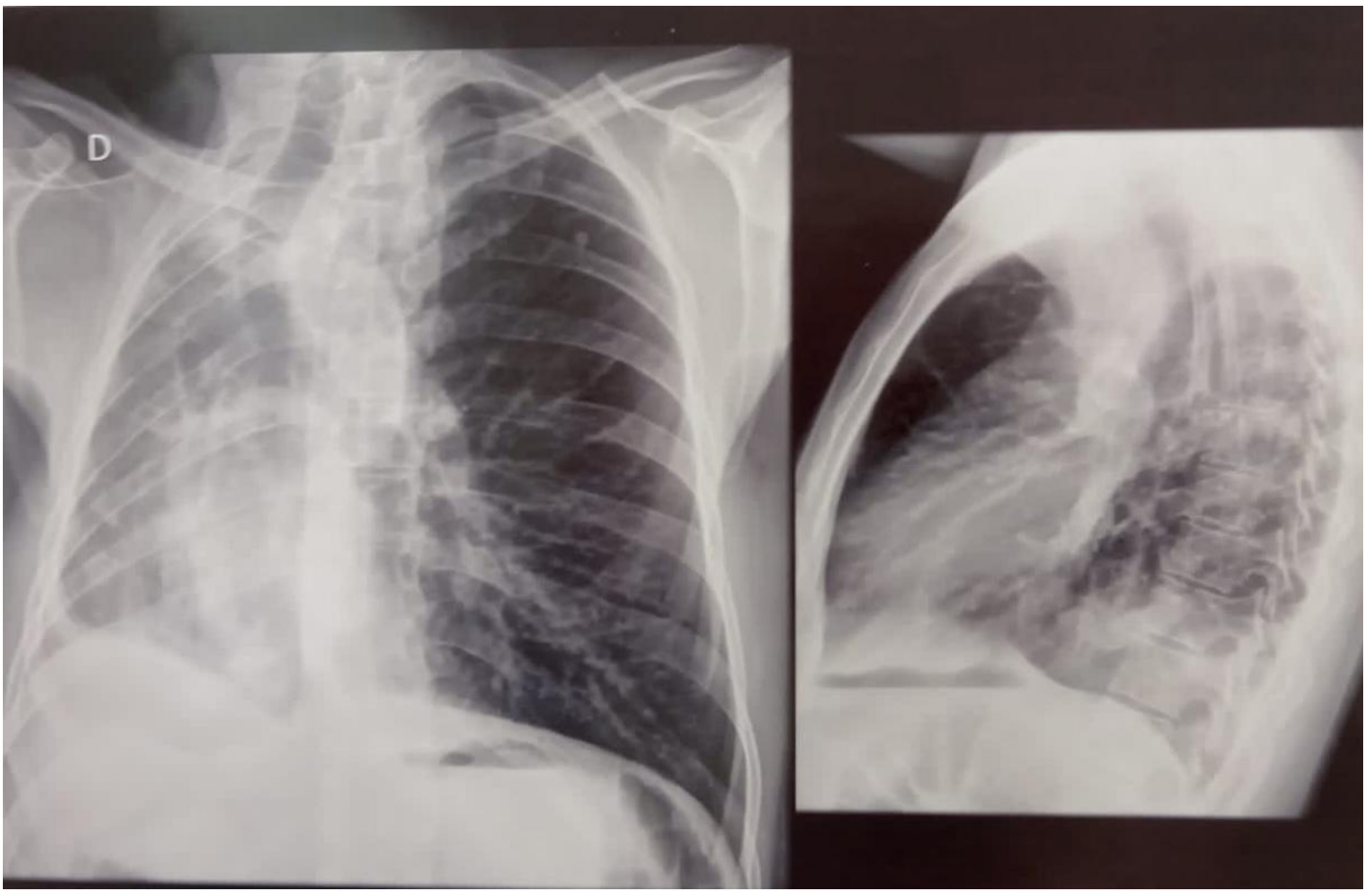

Figure 2. Chest X-ray in PA and lateral views showing right lung agenesis and dextrocardia in the patient.

he had dyslipidemia and anxiety, managed with simvastatin and escitalopram. During the preoperative evaluation, he was classified as ASA II due to his underlying comorbidities. Laboratory tests, including complete blood count and serum electrolytes, were within normal ranges. Spirometry revealed a moderate restrictive ventilatory disorder. Airway assessment identified retrognathia, indicating a potential challenge for intubation. He had previously undergone strabismus correction under general anesthesia with orotracheal intubation without reported complications. Standard intraoperative monitoring included electrocardiography, pulse oximetry, and noninvasive blood pressure measurement. Preoxygenation was performed with 100% oxygen via a closed mask for 3 minutes. Intravenous anesthetic induction was achieved with fentanyl (5 mcg/kg), propofol (3 mg/kg), lidocaine (1 mg/kg), and rocuronium (0.6 mg/kg). Direct laryngoscopy revealed a Cormack-Lehane IIb view, requiring the use of a bougie for successful orotracheal intubation with a 7.5-mm cuffed endotracheal tube. Correct tube placement was confirmed via capnography and auscultation. General anesthesia was maintained with sevoflurane at 1 MAC under pressure-controlled mechanical ventilation. Initial ventilatory parameters included a respiratory rate of 12 breaths per minute, tidal volume of 500 mL, inspiratory pressure of 18 cmH_2O, positive end-expiratory pressure (PEEP) of 5 cmH_2O, and an inspired oxygen fraction (FiO_2) of 49%. After carbon dioxide insufflation for pneumoperitoneum at 11 mmHg, the patient was placed in a head-up position, which led to a decrease in tidal volume to 391 mL while other ventilatory parameters remained stable. Throughout the procedure, hemodynamic stability was maintained, with a mean arterial pressure consistently above 65 mmHg without the need for vasopressor support. Oxygen saturation remained at 100%, heart rate ranged between 80 and 100 beats per minute, and end-tidal CO_2 ($EtCO_2$) was maintained between 30 and 40 mmHg. At the conclusion of the 60-minute procedure, neuromuscular blockade was reversed with 200 mg of sugammadex due to the unavailability of neuromuscular monitoring at the institution. The patient was extubated without complications and exhibited no signs of hemodynamic instability or Fahr's disease-related perioperative events. He recovered uneventfully in the post anesthesia care unit and was discharged the next day with no further complications.

DISCUSSION

Fahr's disease is a rare condition with variable clinical presentations, including movement disorders, cognitive decline, and psychiatric symptoms[2-4]. The pathophysiology involves dysregulated calcium metabolism leading to progressive neurodegeneration[5] From an anesthetic perspective, several concerns arise, including neuromuscular dysfunction: Fahr's disease may affect neuromuscular function due to calcium dysregulation. While there are no established guidelines for neuromuscular blocking agent (NMBA) use in these patients, careful monitoring and reversal are advised to avoid prolonged neuromuscular blockade. Patients with Fahr's disease may be at risk for laryngospasm and dystonic reactions. In this case, no laryngospasm or airway complications occurred during induction and intubation. Although Fahr's disease is not directly associated with cardiac abnormalities, calcium metabolism disturbances may contribute to arrhythmias. Continuous electrocardiographic monitoring is essential. Given the potential for cognitive dysfunction and psychiatric manifestations, anesthetic drugs should be titrated carefully to minimize postoperative delirium or exacerbation of neurological symptoms. To the best of our knowledge there are no previous case reports describing anesthesia in patients with Fahr's disease, apart from a report on anesthesia in Fahr's syndrome, which includes secondary causes of basal ganglia calcification. Dextrocardia requires careful attention to electrocardiographic lead placement to ensure accurate cardiac monitoring. Although this patient remained hemodynamically stable, the literature suggests that transesophageal echocardiography can be beneficial for real-time cardiac function assessment in such cases. Right lung agenesis presents significant challenges in perioperative ventilation, particularly under conditions of increased intra-abdominal pressure during laparoscopic surgery. Pneumoperitoneum can lead to reduced lung compliance, increased airway pressures, and impaired gas exchange. Strategies to optimize ventilation include low tidal volumes: a lung-protective ventilation strategy with lower tidal volumes and adequate PEEP minimizes the risk of barotrauma; PEEP optimization[6]: a PEEP of 5 cmH_2O was applied as part of a lung-protective strategy, which may contribute to minimize the risk of atelectasis in patients with reduced lung volume; capnography monitoring: $EtCO_2$ levels remained stable, suggesting effective ventilation despite lung volume reduction from pneumoperitoneum. A review of existing medical databases (PubMed, LILACS, and MEDLINE) did not identify any established associations between Fahr's disease, dextrocardia, and pulmonary agenesis. This suggests that the co-occurrence of these conditions in this patient is likely a rare coincidence rather than an underlying syndromic association. Postoperative recovery in patients with Fahr's disease should be carefully monitored for neurological deterioration, particularly in response to anesthetic agents or electrolyte imbalances. Fahr's disease has been associated with hypocalcemia and hypomagnesemia, which can influence neuromuscular transmission and the efficacy of NMBAs[5]. Sugammadex allowed rapid and

effective neuromuscular blockade reversal in this case. Patients with right lung agenesis may be at increased risk for postoperative respiratory insufficiency. However, in this case, spontaneous breathing resumed effectively, and the patient had an uneventful postoperative course. Key considerations include careful neuromuscular blockade management, optimized mechanical ventilation strategies, and vigilant cardiovascular monitoring. Despite the rarity of this combination of conditions in this case, the patient's clinical stability and lack of acute neurological manifestations allowed the use of standard anesthetic techniques. Due to the absence of established guidelines and limited published experience, anesthetic care must be individualized based on the patient's functional status and comorbidities.

REFERENCES

1. Gupta B. Anesthetic considerations in a case of Fahr and Primrose syndrome. Res Pract Anesthesiol Open J. 2018;3(1):1-6. https://doi.org/10.17140/RPAOJ-3-116.
2. Jiang W, Mei S, Deng Q, Lei C, Pang A. A case of cerebral large-vessel vasculitis concomitant with Fahr syndrome in systemic lupus erythematosus. Neurologist. 2024;29(1):17-21. https://doi.org/10.1097/NRL.0000000000000520. PMid:37639572.
3. Zhao T, Xu S, Liu S, Xu J, Zhang X, Zhan Y. Fahr's disease linked to a novel MYORG mutation presenting as paroxysmal limb stiffness and dysarthria: case report and literature review. Mol Genet Genomic Med. 2023;11(12):e2276. https://doi.org/10.1002/mgg3.2276. PMid:37680026.
4. Batla A, Tai XY, Schottlaender L, Erro R, Balint B, Bhatia KP. Deconstructing Fahr's disease/syndrome of brain calcification in the era of new genes. Parkinsonism Relat Disord. 2017;37:1-10. https://doi.org/10.1016/j.parkreldis.2016.12.024. PMid:28162874.
5. Park S, Jee DL, Kim H. General anesthesia for a patient with Fahr's syndrome. Medicine (Baltimore). 2019;98(17):e15390. https://doi.org/10.1097/MD.0000000000015390. PMid:31027134.
6. Hedenstierna G. Oxygen and anesthesia: what lung do we deliver to the post-operative ward? Acta Anaesthesiol Scand. 2012;56(6):675-85. https://doi.org/10.1111/j.1399-6576.2012.02689.x. PMid:22471648.

This study was carried out at the Faculdade de Medicina de Jundiaí, Centro de Ensino e Treinamento da Disciplina de Anestesiologia, Jundiaí, São Paulo, Brasil.

Authors' contributions: All authors participated in writing and technical editing of the manuscript.

Ethics statement: Nothing to declare.

Conflict of interest: None.

Financial support: None.

Submitted on: February 19th, 2025
Accepted on: December 5th, 2025

Correspondence
Lucas Lessa Nunes
Faculdade de Medicina de Jundiaí, Centro de Ensino e Treinamento da Disciplina de Anestesiologia
Rua Siracusa 01, 13207-450, Jundiaí, SP, Brasil
Phone: +55 (81) 99323-4589
nunes.lessa@gmail.com