

# Anesthesia During Functional Endoscopic Sinus Surgery for Kartagener's Syndrome: A Case Report and Literature Review

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**Introduction:** Kartagener's syndrome (KS) is a ciliopathic, autosomal recessive disorder characterized by the triad of situs inversus, chronic sinusitis, and bronchiectasis. The abnormal ciliary structure and function lead to variable clinical manifestations, including dextrocardia, pneumonia, bronchitis, chronic rhinosinusitis, otitis media, reduced fertility in women, and infertility in men. This article reports our experience on general anesthesia with endotracheal intubation during functional endoscopic sinus surgery (FESS) in a patient with KS.

**Case Presentation:** A 44-year-old man was admitted to our hospital with chronic nasal obstruction, postnasal drip, chronic sinusitis, and chronic non-productive cough for FESS. The patient's heart was on the right side of his chest. A chest roentgenogram and a high-resolution chest and abdomen computed tomography (CT) scan identified dextrocardia, situs inversus, and chronic bronchitis and bronchiectasis involving both lung bases. CT sinuses showed mucosal thickening of bilateral maxillary and ethmoid and sphenoid sinuses. The patient was prescribed oral medications and nasal spray for crepitations and wheezes heard over bilateral lung fields. Intensive chest physiotherapy and supportive care prior to surgery were provided to prevent worsening of lung function. FESS with bilateral frontal polypectomy was performed. All hemodynamic parameters were stable. The emergence from anesthesia was smooth. After ~20 minutes in the post-anesthesia care unit, the patient was fully awake and pain-free. He was then transferred to the surgical intensive care unit and subsequently to the ward. The postoperative period was uneventful. The patient felt subjectively "very well" and was discharged from the hospital on the 2nd postoperative day.

**Conclusion:** Anesthesiologists must be aware of cardiopulmonary inversion that could challenge the management of patients with KS. To avoid respiratory depression caused by long-acting systemic opioids, we suggest using short-acting opioids during general anesthesia and for postoperative pain relief.

**Keywords:** case report, FESS, general anesthesia, Kartagener's syndrome, sinusitis

## Introduction

Primary ciliary dyskinesia (PCD) is a group of autosomal recessive disorders, and the subgroup that presents with the triad of sinusitis, bronchiectasis, and dextrocardia is called Kartagener's syndrome (KS).<sup>1</sup> Described by Kartagener in 1933,<sup>2</sup> this syndrome has an incidence of 1–2/30,000 birth<sup>3</sup> and is seen in 40% to 50% of patients with PCD.<sup>4</sup>

The ciliary dyskinesia and ineffective mucociliary clearance affect many tissues and organs such as the nasal mucosa and paranasal sinuses. However, the pharynx and the lower respiratory tract down to the respiratory bronchioles are covered by unciliated epithelium.<sup>5</sup>

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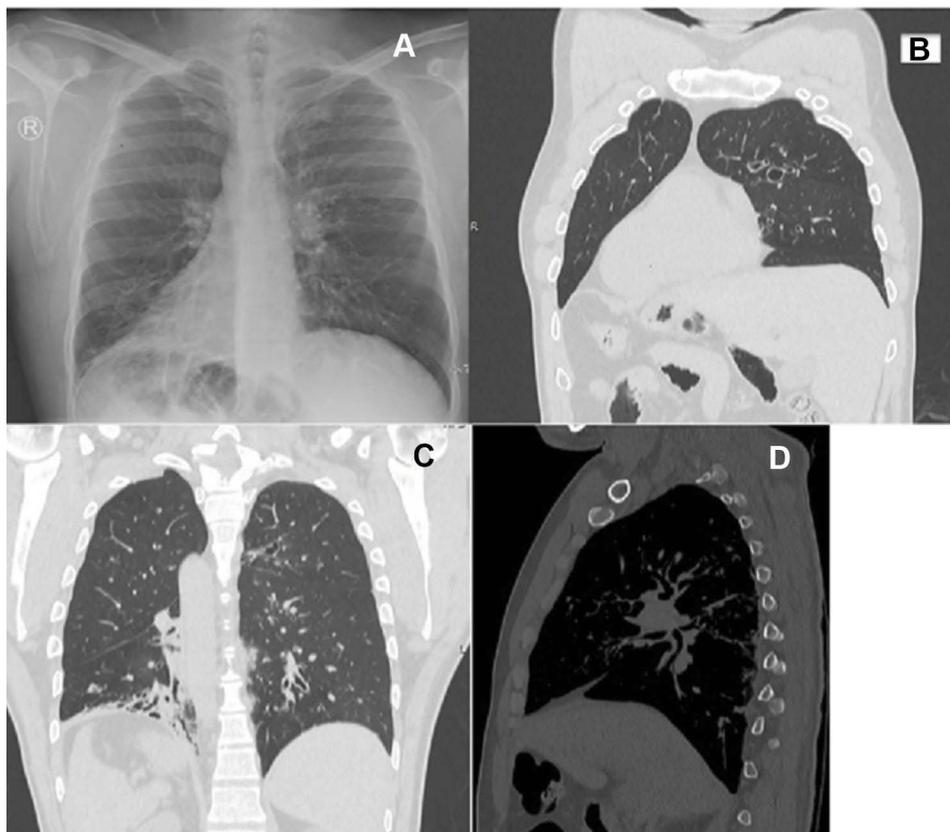
Patients presenting with respiratory distress, chronic pulmonary disease, bronchiectasis, and sinusitis in the setting of KS are a challenge to the anesthesiologist due to the potential respiratory or cardiac complications.<sup>6,7</sup> Therefore, anesthetic management of patients with KS must be conducted carefully. Herein, we describe a case of successful general anesthetic management of a patient with KS who underwent functional endoscopic sinus surgery (FESS).

## Case Presentation

A 44-year-old man was admitted to the Ear, Nose, and Throat department of our hospital in August 2019 with chronic nasal obstruction, postnasal drip, chronic sinusitis, and chronic non-productive cough for FESS. During history-taking, the patient stated that his heart was on the right side of his chest. A chest roentgenogram and a high-resolution chest and abdomen computed tomography (CT) scan identified dextrocardia (Figure 1A), situs inversus (Figure 1B), and chronic bronchitis and bronchiectasis involving both lung bases (Figure 1C and D). CT sinuses showed mucosal thickening of bilateral maxillary and

ethmoid and sphenoid sinuses (Figure 2). KS was diagnosed based on the presence of the classical triad.

On admission, the patient's vital signs, including blood pressure (systolic 112, diastolic 74 mmHg), heart rate (80 beats per minute), respiratory rate (19/min), and the peripheral capillary oxygen saturation (SpO<sub>2</sub>; 94%), were within normal range. The complete blood count and renal and liver function test results were unremarkable. The 12-lead electrocardiogram (ECG) with reversal of the limb and precordial leads showed sinus rhythm and a normal tracing. The patient had a known allergy to acetylsalicylic acid (Aspirin<sup>®</sup>) and nonsteroidal anti-inflammatory drugs, and an allergy panel was performed, which revealed positive hazelnut antibodies. Since crepitations and wheezes were heard over bilateral lung fields, the patient was prescribed oral prednisolone (50 mg, once daily (QD)), oral lansoprazole (30 mg, QD), oral levofloxacin (500 mg, QD), and 0.1% xylometazoline hydrochloride nasal spray (every 8 h). Intensive chest physiotherapy and supportive care prior to surgery were provided to prevent worsening of the lung function. Additional hypertonic saline



**Figure 1** (A) Chest X-ray posteroanterior (PA) view and (B) computed tomography of the chest shows dextrocardia and situs inversus. (C and D) High-resolution computed tomography shows cylindrical bronchiectasis in both lungs.



**Figure 2** High-resolution computed tomography of the sinuses. (A) Shows a mucosal thickening in both maxillary and ethmoid and (B) sphenoid sinuses with diagnosis of chronic sinusitis.

nebulizer (every 8 h) and salbutamol nebulizer (5 mg, twice daily) were provided during pre-anesthetic evaluation in order to prepare the patient for safe anesthesia and surgery. The METs estimated by preoperative history at the preoperative evaluation visit were approximately 4 with moderate-intensity activities. He could walk on a flat surface for one or two blocks and up to two flights of stairs.

In the operating theater, an intravenous (IV) access, neuromuscular monitoring, and routine monitors (ECG, non-invasive blood pressure, and pulse oximetry for SpO<sub>2</sub>) were established. The patient was pre-oxygenated with a face mask for 3 minutes. General anesthesia was induced with 2 µg/kg IV fentanyl and 2–2.5 mg/kg IV propofol. Neuromuscular blockade was achieved with 0.5 mg/kg atracurium. The trachea was intubated in a single attempt using a single-lumen cuffed tube of internal diameter 8.0 mm. The anesthesia was maintained with 50% O<sub>2</sub>, N<sub>2</sub>O, and isoflurane. Remifentanyl was given continuously with a syringe infusion pump (Perfusor® Space; B. Braun, Germany) at a rate of 0.1 µg/kg/min adjusted per the patient's heart rate.

The patient's lungs were ventilated with a 50% fraction of inspired oxygen. A tidal volume of 7–10 mL/kg and respiratory rate of 10–12 breaths/min were adjusted to maintain the End-tidal CO<sub>2</sub> between 30 and 36 mmHg. To increase the lung functional residual capacity a small amount of PEEP (5 cmH<sub>2</sub>O) was applied during the mechanical ventilation of the patient. A single dose of IV cefotaxime 1 g was administered intraoperatively.

About 20 minutes before the end of the surgery, 1 g paracetamol with 100mg tramadol was infused for 15 minutes. Dexamethasone 8 mg and metoclopramide 10 g were also given intravenously.

The duration of FESS with bilateral frontal polypectomy was 95 minutes. All hemodynamic parameters were stable, and no additional muscle relaxant or fentanyl was required throughout the procedure. At the end of the surgery, the residual neuromuscular block was reversed with IV neostigmine 0.05 mg/kg and IV atropine 1 mg.

Lungs suctioning through the endotracheal tube and oral suctioning were performed gently. Tracheal extubation was performed when the patient was able to open the eyes and ventilate spontaneously. The emergence from anesthesia was smooth and without complications. After ~20 minutes in the post-anesthesia care unit, the patient was fully awake and pain-free (pain scale 2). The patient was then transferred to the surgical intensive care unit for 8 hours, and subsequently to the ward. An arterial blood gas analysis on room air showed pH = 7.419, oxygen concentration (PaO<sub>2</sub>) = 70 mm Hg, carbon dioxide concentration (PaCO<sub>2</sub>) = 36 mm Hg, bicarbonate ion concentration (HCO<sub>3</sub><sup>-</sup>) = 23 mmol/L, O<sub>2</sub> saturation = 94%, and base excess = -1 mmol/L. The postoperative period was uneventful. The patient felt subjectively "very well" and was discharged from the hospital on the second postoperative day.

## Discussion and Conclusion

KS is the most common type of PCD, characterized by chronic recurrent rhinosinusitis, bronchiectasis, and situs inversus. As a rare autosomal recessive inherited disease, its incidence is estimated to be 1 in 32,000.<sup>2</sup> Dysfunction of the motile cilia causes ineffective mucus clearance and organ laterality defects. Affected patients usually present with chronic infections of the upper and lower respiratory tract, otitis media, pneumonia, and bronchiectasis, often complicated by infections with *Pseudomonas* spp.<sup>8,9</sup>

Impaired ciliary motility could cause perioperative respiratory difficulty in patients with KS.<sup>10</sup> Serapinas et al<sup>11</sup> recommend a combination of physiotherapy and physical exercise to enhance respiratory tract clearance, along with the aggressive treatment of upper and lower respiratory tract infections. We provided intensive physiotherapy and supportive pulmonary care prior to surgery for our patient, along with the antibiotic levofloxacin for preventing the progressing of the respiratory tract infection. Appropriate preoperative respiratory tract management is also essential to prevent potential peri- and postoperative morbidities such as acute respiratory failure and pulmonary edema.<sup>12</sup>

General anesthesia or regional techniques such as neuraxial anesthesia could be chosen for patients with KS, with the regional techniques presenting fewer risks than general anesthesia. In our patient, we used propofol for the induction of anesthesia. Propofol dilates the bronchi, slightly decreases mucus production, and suppresses the airway reflexes.<sup>10</sup> Wu et al<sup>13</sup> report significantly better protection against an increase in airway resistance using propofol than that using thiopentone after tracheal intubation. To achieve muscle relaxation for facilitating endotracheal intubation, we chose atracurium for our patient, primarily due to its ability to degrade spontaneously in plasma and tissues at normal body pH and temperature (Hofmann degradation). We maintained anesthesia with 1–1.3 minimum alveolar concentration of isoflurane as inhalational anesthetics cause bronchodilation in a dose-related manner.<sup>13</sup> This is especially important in patients with a compromised respiratory tract due to KS.

Dextrocardia has an incidence rate of ~2 per 10,000 births. Associated congenital cardiac anomalies occur in less than 3% with dextrocardia and include inter alia, atrial septal defects, ventricular septal defects, and pulmonary valve stenosis. When ECG leads are reversed, patients with dextrocardia show sinus rhythm and the heart mostly appears normal without other abnormalities, as seen in our patient.<sup>14,15</sup>

In conclusion, anesthesiologists must be aware of cardiopulmonary inversion that could challenge the pre-, peri-, and postoperative management of patients with KS. To avoid the respiratory depression caused by long-acting systemic opioids, we suggest using short-acting opioids during general anesthesia and for postoperative pain relief. Cardiopulmonary complications could be prevented to a large extent by proper perioperative suctioning of the pharynx and pulmonary tract and adequate postoperative management to prevent nausea and vomiting.

## Abbreviations

CT, Computed tomography; FESS, functional endoscopic sinus surgery; KS, Kartagener's syndrome; PCD, primary ciliary dyskinesia; QD, once daily; ECG, electrocardiogram; IV, intravenous.

## Data Sharing Statement

The data that support the findings of this study are available on request from the corresponding author, KEL.

## Ethics and Consent Statement

Based on the regulations of the department of research of the Jordan University of Science and Technology, an institutional review board approval is not required for case reports.

## Consent for Publication

A written permission for the use of patient data for publication was obtained.

## Author Contributions

All authors made substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; took part in drafting the article or revising it critically for important intellectual content; gave final approval of the version to be published; and agree to be accountable for all aspects of the work.

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## Disclosure

The authors declare that they have no competing interests.

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