

KARTAGENER SYNDROME AND ANESTHETIC IMPLICATIONS: PATHOPHYSIOLOGICAL REVIEW AND CASE REPORT IN ANTERIOR CERVICAL ARTHRODESIS

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ABSTRACT

Primary ciliary dyskinesia (PCD) is an inherited disorder in which motile cilia have structural and/or functional defects, impairing mucociliary clearance and promoting secretion retention, bacterial colonization, recurrent infections, and chronic inflammation from childhood, with potential progression to bronchiectasis and respiratory failure. Kartagener syndrome (KS) is the classic form associated with PCD, combining chronic respiratory disease with laterality defects such as situs inversus totalis or dextrocardia, which affects the interpretation of ECG, auscultation findings, and imaging studies. Diagnosis is multimodal (clinical features plus specialized tests), and management aims to relieve symptoms and prevent exacerbations (vaccination, respiratory physiotherapy, and airway clearance). We report the case of a 50-year-old patient with KS undergoing anterior cervical fusion, clinically stable and without recent exacerbation. Total intravenous anesthesia (TIVA) with propofol/remifentanyl was performed, with rocuronium titrated under train-of-four (TOF) monitoring and depth-of-anesthesia monitoring, alongside lung-protective ventilation and periodic recruitment maneuvers. The course was uneventful, and extubation was performed under strict criteria after complete neuromuscular blockade reversal to ensure effective cough and reduce the risk of mucus plugging/atelectasis. The discussion emphasizes that perioperative complications in KS are predominantly respiratory (atelectasis, bronchospasm, infection) and highlights humidification, secretion management, lung-protective ventilation, complete neuromuscular reversal, adequate analgesia, and early respiratory physiotherapy as key safety measures.

Keywords: Kartagener syndrome, Ciliary motility disorders, Anesthesia, Rare diseases, Dextrocardia, Situs inversus.

INTRODUCTION

Primary ciliary dyskinesia (PCD) is a hereditary disease, predominantly autosomal recessive, in which motile cilia present structural or functional abnormalities. Because

these cilia are essential for mucociliary clearance, impairment of their movement reduces airway clearance and facilitates the accumulation of secretions in the upper and lower respiratory tract. In practice, this favors bacterial colonization, recurrent respiratory infections, and chronic inflammation. For this reason, the clinical picture usually begins early in life, often during childhood, with chronic rhinosinusitis, recurrent otitis, and frequent episodes of bronchitis or pneumonia. Over the years, persistent inflammation may lead to bronchiectasis and recurrent exacerbations, and in more advanced cases it may progress to chronic respiratory failure, pulmonary hypertension, and cor pulmonale.¹

Kartagener syndrome (KS) is the classic presentation associated with PCD and is characterized by the combination of chronic respiratory disease with laterality abnormalities. Among these alterations, situs inversus totalis and dextrocardia stand out. Situs inversus totalis refers to the “mirror-image” inversion of the position of thoracic and abdominal organs, involving the heart and viscera. Dextrocardia specifically describes the heart located in the right hemithorax, with the apex pointing to the right. Dextrocardia may occur as part of situs inversus totalis, but it can also occur in isolation, which requires additional attention during anatomical and cardiological evaluation. Although a significant proportion of patients with PCD present situs inversus, KS should be understood as a spectrum, because the triad is not always present in its complete form, which may delay clinical recognition.¹

Historically, Kartagener described the clinical triad in 1933, and later the understanding of the syndrome advanced with the identification of cilia axonemal abnormalities, such as the absence of dynein arms, as well as the association with infertility, particularly in males.^{2,3}

The diagnosis of PCD and KS is challenging because there is no single test considered a gold standard in all contexts. Current recommendations emphasize integrating the clinical picture with specialized complementary tests, such as nasal nitric oxide measurement, high-speed video analysis of ciliary beating, transmission electron microscopy, immunofluorescence, and genetic testing, depending on the availability and expertise of the center. Guidelines from the European Respiratory Society (ERS) and the American Thoracic Society (ATS) reinforce this multimodal diagnostic reasoning and the classification of diagnosis based on probability.^{4,5}

Because there is no specific curative treatment, management is aimed at reducing symptoms, preventing exacerbations, and limiting the progression of bronchial damage. This strategy includes vaccination, respiratory physiotherapy, and bronchial hygiene, in addition to early treatment of exacerbations and measures directed at chronic colonization when indicated.^{1,5}

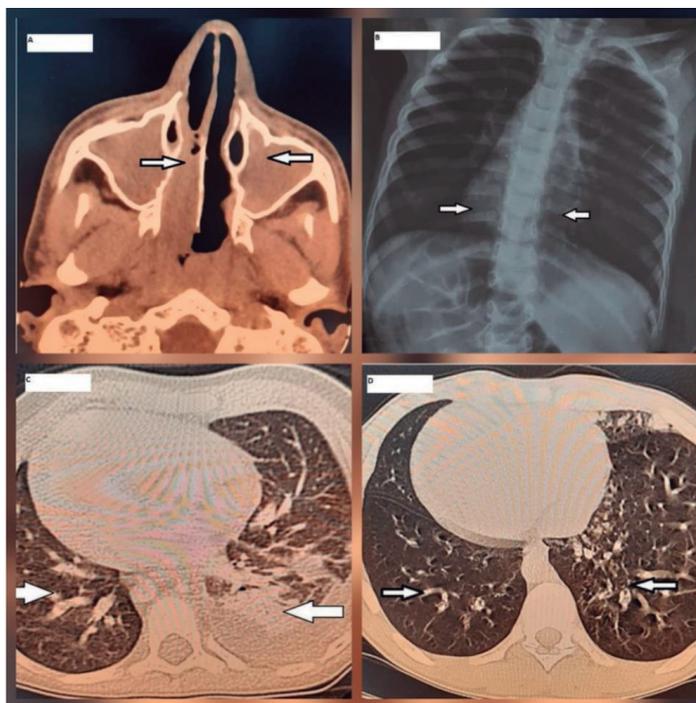


Figure 1. Illustrative images of Kartagener Syndrome. A) CT scan showing chronic sinusitis with nasosinusual polyposis. B) Chest X-ray showing a case of dextrocardia with findings of pneumonia and minimal pleural effusion in the left upper lobe. C) Thoracic CT demonstrating situs inversus, bronchial dilation, and left laterobasal emphysema. D) Chest CT demonstrating bronchial dilation associated with situs inversus.⁶

CASE REPORT

A 50-year-old patient with KS underwent anterior cervical arthrodesis. In the immediate preoperative period, she was stable, without signs of recent respiratory exacerbation and without hypersecretion on the day of the procedure; therefore, no additional tests were requested at that time. Nevertheless, considering the baseline risk of secretion retention and atelectasis during controlled ventilation, anesthetic management was planned with a focus on ventilatory stability and prevention of pulmonary complications.

TIVA was performed using target-controlled infusion of propofol and remifentanyl, associated with rocuronium, with neuromuscular blockade titration guided by train-of-four (TOF) monitoring. The anesthetic plan was monitored with SedLine, a processed electroencephalogram monitor used to estimate hypnotic depth, with a purpose similar to BIS, assisting in the titration of the anesthetic depth. The airway was managed with orotracheal intubation, and during maintenance protective ventilation was used, with periodic alveolar recruitment maneuvers, aiming to reduce alveolar collapse and maintain an adequate ventilation–perfusion relationship.

The intraoperative period was uneventful, and at the end of the procedure the patient presented a smooth emergence, without bronchospasm and without hypoxemia. Considering that the anterior cervical approach may be associated with local edema and, more rarely,

hematoma with potential airway compromise, extubation was performed using strict criteria, prioritizing complete ventilatory recovery and full reversal of neuromuscular blockade. In KS, this precaution is particularly relevant, as effective cough and full spontaneous ventilation reduce the risk of mucous plugging and postoperative atelectasis.

DISCUSSION

The pathophysiological basis of KS can be understood through a relatively simple cycle: retained secretions favor infection, infection fuels inflammation, and inflammation, over time, promotes remodeling and structural damage. The failure of mucociliary clearance leaves secretions more stagnant and difficult to remove, creating an environment conducive to recurrent infections and perpetuation of the inflammatory process. This mechanism explains why bronchiectasis and chronic sinus disease are so common, in addition to the risk of progressive loss of functional respiratory reserve in some patients.³

Laterality abnormalities also have practical implications in the perioperative setting. When dextrocardia is present, for example, a standard electrocardiogram (ECG) lead placement may suggest abnormalities that actually reflect the “mirror-image” anatomy. The same reasoning applies to the location of auscultation foci, interpretation of imaging studies, and some decisions regarding equipment positioning. Therefore, identifying in advance whether the patient has situs inversus totalis or isolated dextrocardia helps avoid confusion and improves perioperative planning.^{1,4,5}

In anesthesia, the central issue is the increased respiratory risk. Impaired mucociliary clearance favors mucous plugs and atelectasis during mechanical ventilation. In addition, chronic inflammation may be associated with bronchial hyperreactivity, increasing the risk of bronchospasm and hypoxemia during airway manipulation. In patients with bronchiectasis and bacterial colonization, the risk of respiratory infection and postoperative exacerbation is also higher. A systematic review of published cases shows that respiratory complications account for most of the perioperative events described in KS.⁶

In practice, this means that preoperative evaluation is guided by the patient’s current clinical status. If there are signs of recent exacerbation, such as fever, increased sputum production, wheezing, or worsening dyspnea, the usual approach is to optimize bronchial hygiene and respiratory physiotherapy before the procedure and consider additional evaluation depending on severity. When there is no exacerbation, management focuses on the prevention of intraoperative and postoperative complications.^{5,7}

During general anesthesia, some measures are particularly consistent with the underlying pathophysiology. Humidification of inspired gases helps prevent secretions from becoming thicker and more difficult to mobilize. Protective ventilation strategies, with alveolar recruitment maneuvers adjusted to the clinical context, help reduce atelectasis. Airway suctioning should be performed judiciously, with particular attention at the end of the procedure, avoiding mucous plugs and minimizing airway trauma. Postoperatively, the focus is on restoring full spontaneous ventilation and effective coughing, as these help compensate, at least partially, for impaired mucociliary clearance.⁷

At this point, complete reversal of neuromuscular blockade is essential. Monitoring with train-of-four (TOF) allows safer titration and reduces the risk of residual neuromuscular

blockade, which is particularly relevant in KS because effective coughing and secretion mobilization are key to preventing atelectasis and mucus retention in the postoperative period. Adequate analgesia and early respiratory physiotherapy complete this strategy, as they facilitate deep ventilation, pulmonary expansion, and bronchial hygiene.⁷

It is also pertinent to consider the impact of anesthetic technique on mucociliary transport. A clinical trial demonstrated worse mucus transport with inhalational anesthesia compared with total intravenous anesthesia (TIVA) using propofol and remifentanyl.⁸ In parallel, experimental studies suggest that propofol may modulate ciliary motility through the nitric oxide (NO) and cyclic guanosine monophosphate (cGMP) pathway, a mechanism that supports the biological plausibility of this choice in patients with impaired mucociliary clearance.⁹ Taken together, reviews and case reports describe TIVA as an appropriate strategy in KS, provided it is combined with consistent measures for secretion management and prevention of respiratory complications.^{7,10,11}

In situations requiring selective intubation, such as thoracic surgery, situs inversus may require specific planning and careful confirmation of device positioning, ideally with bronchoscopy when available, to reduce the risk of inadequate ventilation.¹²

CONCLUSION

Kartagener syndrome is a condition associated with primary ciliary dyskinesia (PCD), characterized by chronic respiratory disease and laterality abnormalities in some cases. In the perioperative setting, the risk is mainly related to respiratory complications, particularly atelectasis, bronchospasm, and infection, in addition to specific considerations in the interpretation of diagnostic tests when dextrocardia and, especially, situs inversus totalis are present. Safe anesthetic management depends on recognizing and optimizing exacerbations, carefully managing secretions, ensuring adequate humidification, applying protective ventilation strategies, and performing extubation only after complete recovery from neuromuscular blockade, in addition to effective analgesia and early respiratory physiotherapy. In the presented case, the strategy using TIVA, protective ventilation, and appropriate monitoring was consistent with the pathophysiology of KS and the available literature, contributing to a favorable perioperative outcome.

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