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INVERSION OF BRONCHIAL ANATOMY AND LUNG SEGMENTS IN KARTAGENER SYNDROME: A RETROSPECTIVE ANALYSIS OF FIVE SITUS INVERSUS CASES AND THEIR CLINICAL IMPLICATIONS

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ABSTRACT

Kartagener's Syndrome (KS), a subset of primary ciliary dyskinesia, is characterized by a triad of chronic sinusitis, bronchiectasis, and situs inversus. This retrospective analysis examines cases of five patients diagnosed with situs inversus as part of KS, focusing on the inversion of bronchial anatomy and lung segments. Detailed imaging assessments revealed a consistent mirroring of bronchial tree structures and lung segments, necessitating a tailored approach for diagnostic and procedural strategies. The findings highlight the importance of recognizing altered anatomical landmarks to prevent procedural complications and improve patient outcomes. Furthermore, this article discusses how awareness of these unique presentations can inform targeted therapies and surgical planning, ultimately enhancing the quality of care for patients with KS. Knowledge of these clinical implications is essential for healthcare providers managing this rare congenital disorder.

Article

Kartagener's syndrome, a form of primary ciliary dyskinesia (PCD), is a rare congenital genetic disorder with an incidence rate of 1 in 32,000 live births.[1] The incidence is notably higher among populations with a prevalence of consanguineous marriages.[2] This syndrome is also called Kartagener's triad, Afzelius syndrome, Siewert's syndrome, or the dextrocardia-bronchiectasis-sinusitis syndrome.

Kartagener's syndrome is inherited in an autosomal recessive manner.[3,4] It is primarily characterized by three clinical features: bronchiectasis, chronic sinusitis, and situs inversus, the latter being the transposition of the visceral organs.[3,4] The disorder stems from gene mutations encoding the dynein protein, a crucial component of cilia.[5,6] These genetic mutations impair ciliary function or motility during organogenesis and in fully developed organs, leading to manifestations such as situs inversus, infertility, azoospermia, and recurrent sinus and respiratory infections.

Impairments in ciliary beating result in the erratic rotation of primitive organ precursors, contributing to the development of situs inversus. Structural abnormalities of the dynein arm are present in approximately 90% of PCD patients.[7] The principal genetic mutations associated with PCD involve the dynein genes DNAI1 and DNAH5.[8] Approximately half of the individuals affected by PCD exhibit situs inversus and are diagnosed with Kartagener's syndrome.[9]

The diagnosis of Kartagener's syndrome relies on both clinical and radiological evaluations. Primary ciliary dyskinesia (PCD), however, can be confirmed through a series of screening and diagnostic tests, including the measurement of exhaled nitric oxide, the saccharin test, and nasal mucosa biopsy for ciliary beat pattern and frequency analysis through video recording. Additionally, electron microscopy can identify ultrastructural defects in the cilia.[10] The impaired ciliary function results in deficient mucociliary clearance in the airways and sinuses, leading to recurrent infections and subsequent damage to affected organs. Bronchiectasis and chronic sinusitis represent significant clinical consequences of Kartagener's syndrome, necessitating medical intervention.

This study reviewed radiological images of the lungs in five cases of Kartagener's syndrome to examine the central airway patterns. Due to limited access to advanced diagnostic procedures, all cases were diagnosed based on clinical-radiological criteria. When situs inversus is considered in clinical practice, attention is primarily directed towards unilateral organs such as the heart, liver, stomach, pancreas, and spleen, often

neglecting the inversion of lung segments and bronchial anatomy.

In individuals with Kartagener's syndrome, the bronchial tree, including the trachea and branching bronchi, is typically reversed or mirrored from its normal anatomical position [Fig 1]. Specifically, the trachea and main bronchi are located on the left side of the chest, with the left lung possessing three lobes and the right lung two lobes. These anatomical variations pose significant clinical challenges during specific bronchial procedures.

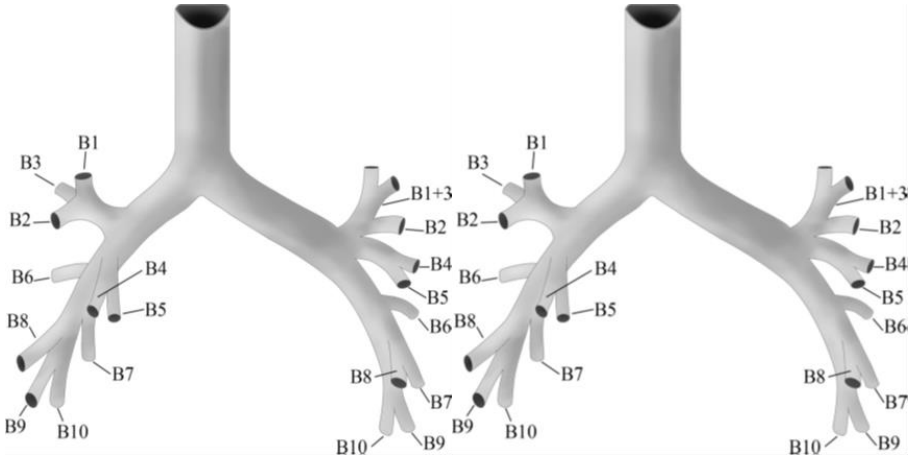
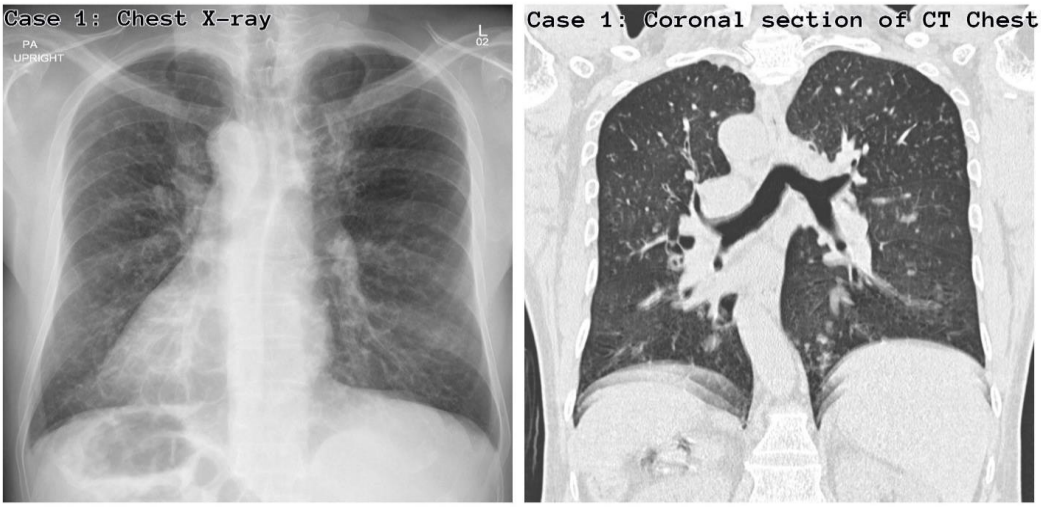


Fig. 1: Drawing illustrates the segmental bronchial anatomy with its mirror image (observed in Kartagener syndrome) according to Boyden's nomenclature. On the right side, B1 = apical bronchus, B2 = anterior bronchus, B3 = posterior bronchus, B4 = lateral bronchus, B5 = medial bronchus, B6 = superior bronchus, B7 = medial basal bronchus, B8 = anterior basal bronchus, B9 = lateral basal bronchus, and B10 = posterior basal bronchus. On the left side, B1 and B3 arise from the same trunk and form the apicoposterior bronchus (B1 + 3), B4 = superior lingular bronchus, and B5 = inferior lingular bronchus. The other left segmental bronchi have the same designations as their right-sided counterparts.[11]

This study observed that all five cases presented with bronchiectasis, chronic sinusitis, and situs inversus. Notably, the bronchial anatomy of these patients was inverted. The left bronchial tree typically exhibited a short, wide-diameter left main bronchus. In these instances, the left upper lobe bronchus originated independently from the left main bronchus, deviating from the usual pattern of merging with the lingular segment bronchi to form the upper division. Additionally, the left lower bronchial tree resembled an intermediate bronchus rather than a lower division, subsequently branching into what would be equivalent to the left-sided middle lobe and lower lobe bronchi.

Conversely, the right bronchial tree in these cases demonstrated a long, narrow-diameter right main bronchus, with the right upper lobe bronchus originating as the upper division of the bronchial tree. It also showed an analogue to the right-sided lingular segment. Across all five cases, this anatomical inversion suggested the presence of left lung anatomy within the right hemithorax and vice versa.



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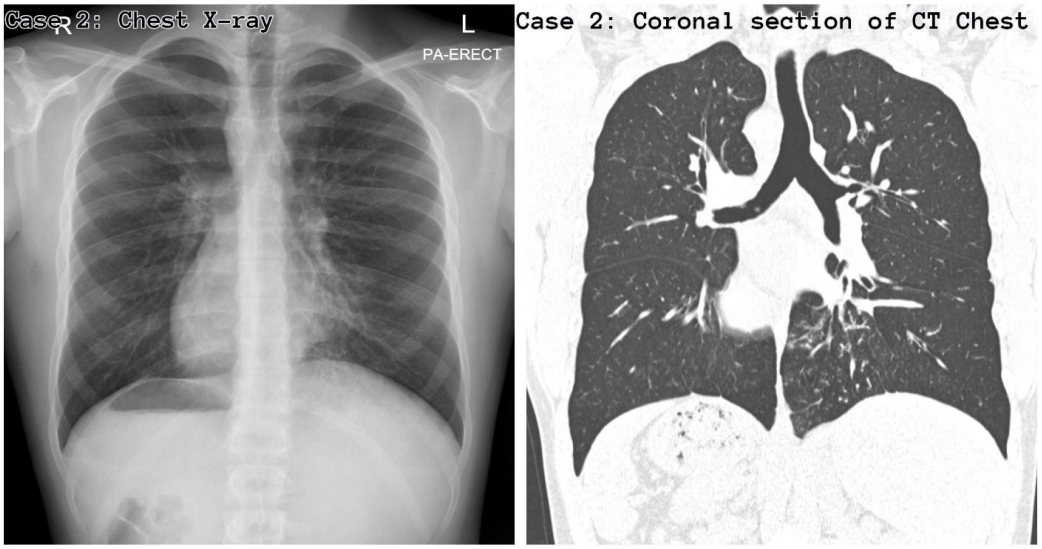

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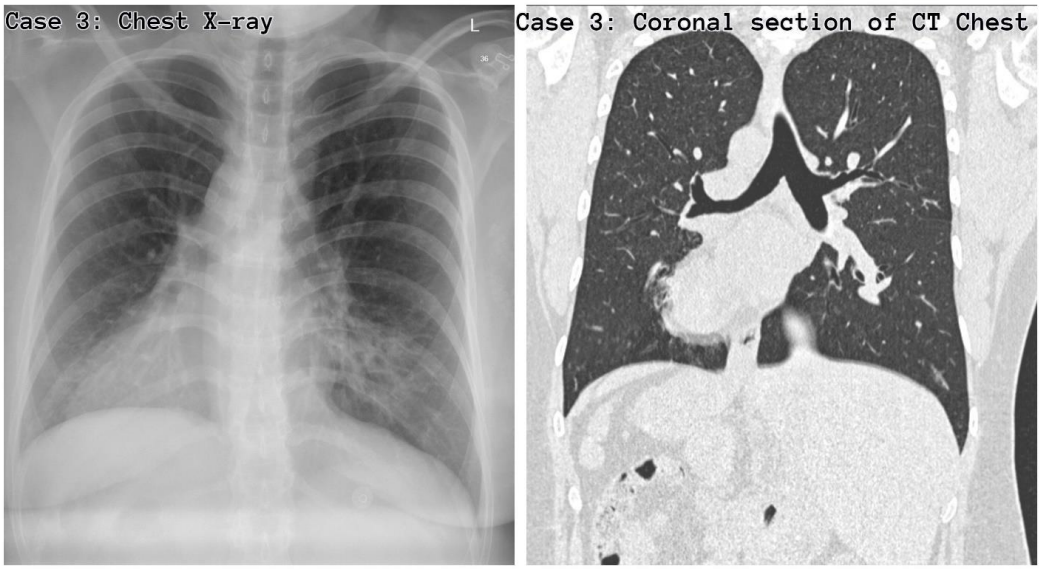

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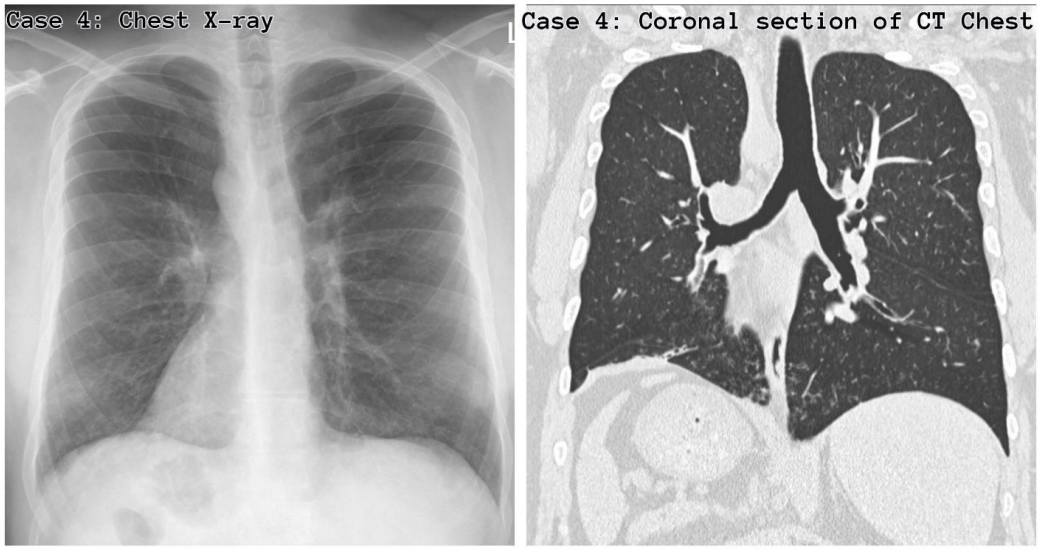

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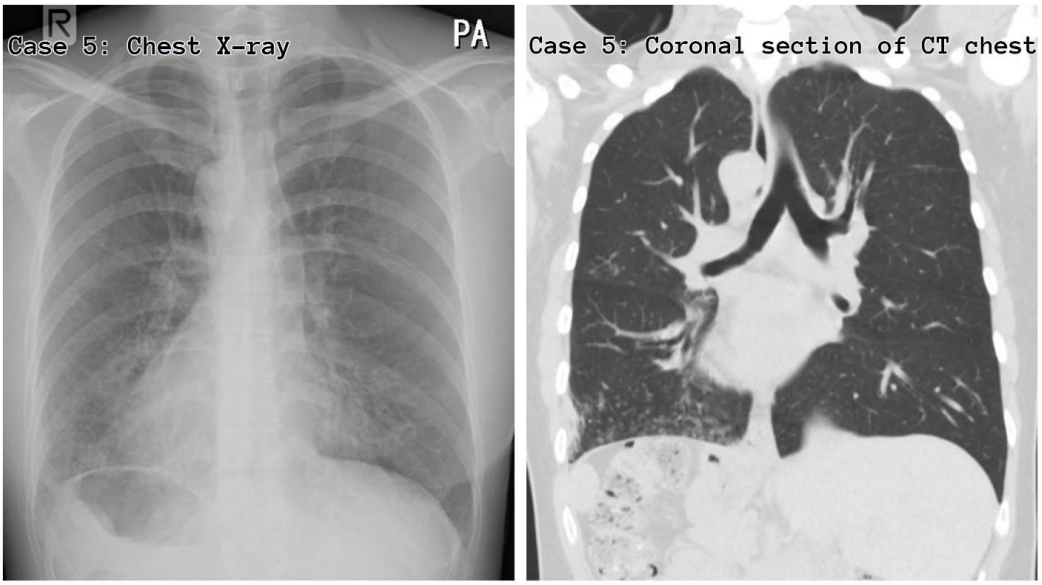

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The inversion of bronchial anatomy holds no specific clinical significance. However, the mirror-image configuration of the bronchi complicates lung isolation. Fibreoptic bronchoscopy may be required to evaluate the bronchial tree for one-lung ventilation, as a comprehensive understanding of bronchial anatomy is essential in such cases. The mirrored airway anatomy necessitates meticulous planning for the placement of lung separation devices. One-lung ventilation may be indicated to confine infection or bleeding to a single lung for lung resection, repair of thoracic aortic aneurysm, oesophagal surgery, whole lung lavage, or thoracoscopy.[12,13,14]

When single-lung ventilation is required, anesthesiologists generally avoid deep intubation of the right main bronchus because the endotracheal tube can obliterate the right upper lobe bronchus due to its short segment. In Kartagener's syndrome, this adverse effect can similarly occur with left-sided intubation.

For double-lumen endotracheal tube (DLT) intubation, left-sided DLTs are typically preferred as they are easier to position without obstructing the upper lobe bronchus. A right-sided DLT includes an orifice along the bronchial tube to allow ventilation of the right upper lobe. However, right-sided DLTs are challenging to place without obstructing the right upper lobe bronchus. In patients with Kartagener syndrome, a left-sided DLT with the bronchial lumen oriented toward the right side is placed into the anatomical left main bronchus located in the right hemithorax.[15]

CONCLUSION

In conclusion, although the inversion of the bronchial tree in cases of situs inversus, particularly in Kartagener's syndrome, is not extensively documented in medical literature, it remains an essential anatomical variation that must be acknowledged. This awareness is especially critical for anesthesiologists during procedures like single-lung ventilation or double-lumen tube intubation, as the inversion can present significant challenges, particularly when intubating the left bronchus. Additionally, the increased risk of foreign body aspiration into the left lung in Kartagener's cases further emphasizes the need for healthcare providers to consider these unique anatomical features carefully. Understanding and anticipating these variations can help prevent potential complications and optimize patient care during treatment and interventions.

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