International Clinical and Medical Case Reports Journal
Case Report (ISSN: 2832-5788)

# Ventilator Associated Pneumonia in Patient with Tracheobronchomegaly (Mounier-Kuhn Syndrome) and Hereditary Angioedema

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**Citation:** *Kahn D, Khan M, Sposato A, Yalcin Y, Devi A, Bajpayee L, et al. Ventilator Associated Pneumonia in Patient with Tracheobronchomegaly (Mounier-Kuhn Syndrome) and Hereditary Angioedema. Int Clinc Med Case Rep Jour. 2024;3(4):1-10.*

**Received Date:** 07 April, 2024**; Accepted Date: :** 12 April, 2024**; Published Date: :** 15 April, 2024

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## INTRODUCTION

Mounier-Kuhn syndrome (MKS) is a rare congenital disorder characterized by a significant dilatation of the trachea and main bronchi, which can result in bronchiectasis. It manifests with recurrent lower respiratory tract infections due to aspiration. These patients are at a high risk of intubation and consequently ventilator associated pneumonia due to difficult peri-intubation management.

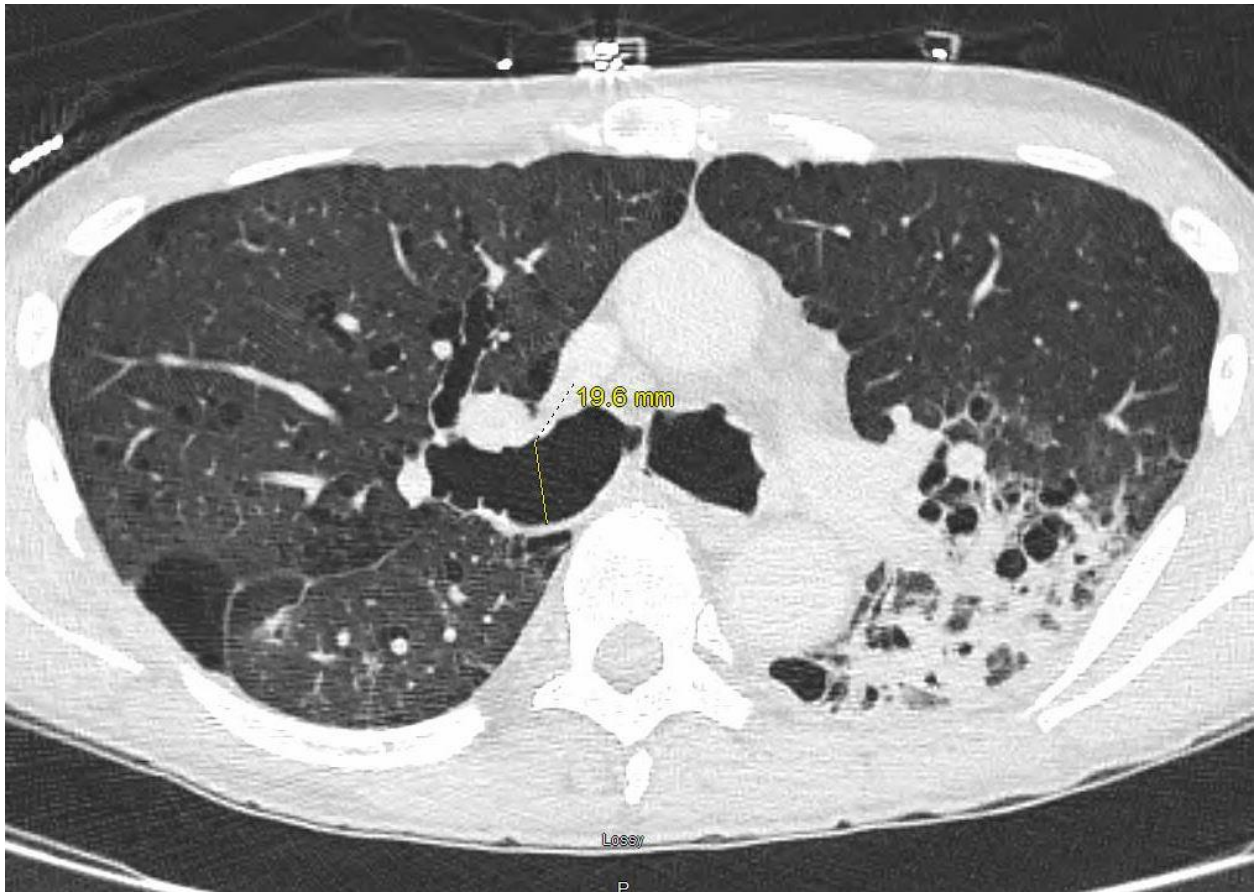
## CASE PRESENTATION

The patient is a 58-year-old man with PMH of AUD, asthma, hypertension, right sided Bell's Palsy who presented to ED with a sore throat and difficulty speaking, which had been progressing for two days prior to presentation. On arrival to the ED, the patient was afebrile, tachycardic to the 110's, and saturating well on room air. On exam, the patient had oropharyngeal swelling and was unable to speak or handle secretions, and was diagnosed with angioedema presumably secondary to ACEi. A CT of the neck was

conducted and incidentally found tracheobronchomegaly (3.6cm) associated with multiple large diverticula, multiple tracheal cartilage destruction, dilatation of main bronchus, global emphysema, and ground glass opacities with a tree-in-bud appearance within the right upper lobe compatible with aspiration. The patient was urgently intubated for airway protection. The patient was successfully extubated on the third day of admission. On the fourth day of admission, the patient developed a fever. At that time, a chest X-ray showed a new airspace opacity in the left mid to lower lung zone, likely representing a large volume aspiration pneumonia, and the patient was treated with ampicillin/sulbactam. He was subsequently discharged to a skilled nursing facility with close follow up.

## DISCUSSION

MKS is a rare disorder with only several hundred reported cases. The etiology is uncertain but is related to the absence of smooth muscle and elastic connective tissue in the trachea and main bronchi. The clinical picture varies and usually includes nonspecific respiratory symptoms, such as cough, dyspnea, and recurrent airway infection[1]. Radiological criteria for MKS include dilatation of trachea >3cm, right mainstem bronchus dilatation >2.4cm, and left mainstem bronchus >2.3cm[2]. In MKS, intubation can be challenging and can lead to multiple complications, such as peritubular air leakage, leading to aspiration and inadequate ventilation, and even fatal rupture of diverticula. Thus, the appropriate type and size of endotracheal tube and appropriate ventilator settings must be chosen carefully[3]. Although no studies have been conducted on the subject, one might consider empiric antibiotic treatment in ventilated patients with MKS due to a very high risk of aspiration due to an unfitted ETT cuff, as seen in our reference patient[1-3] (Figure 1).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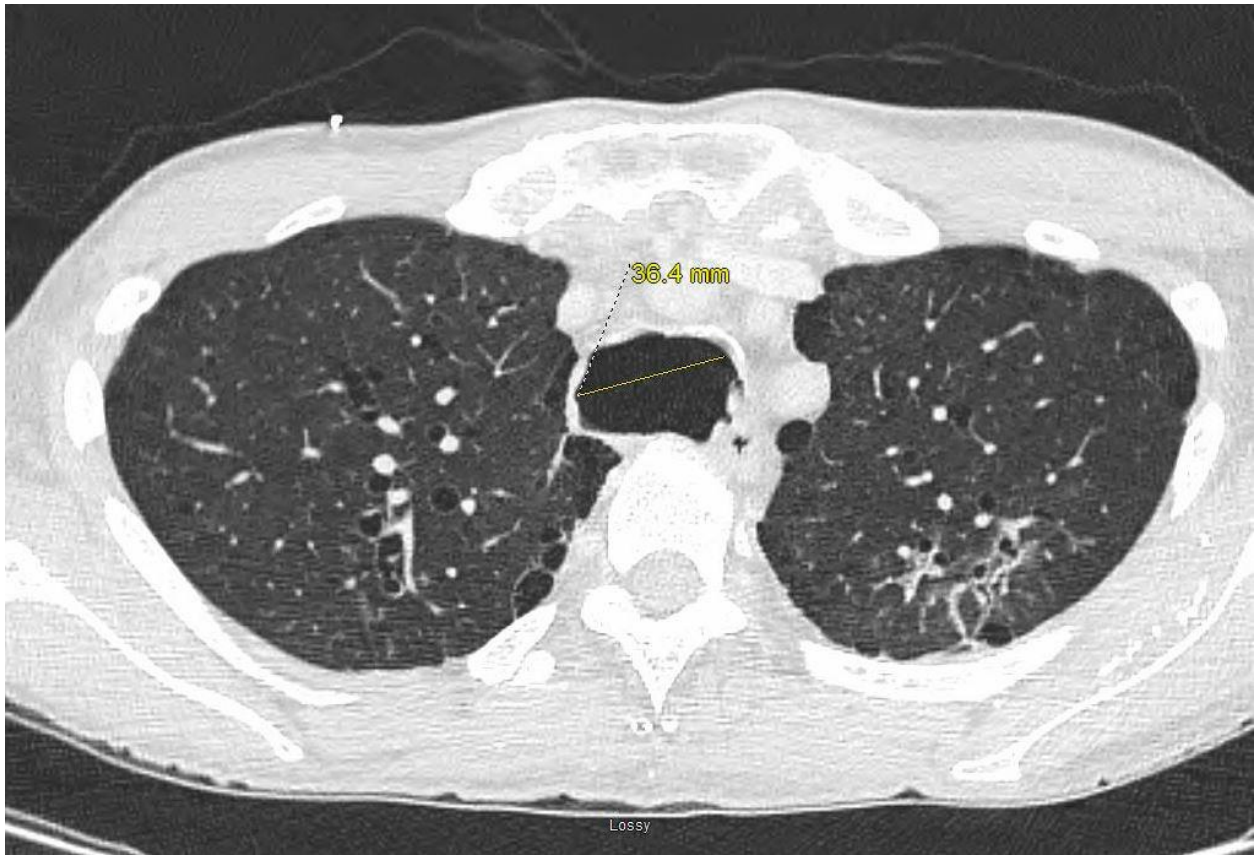
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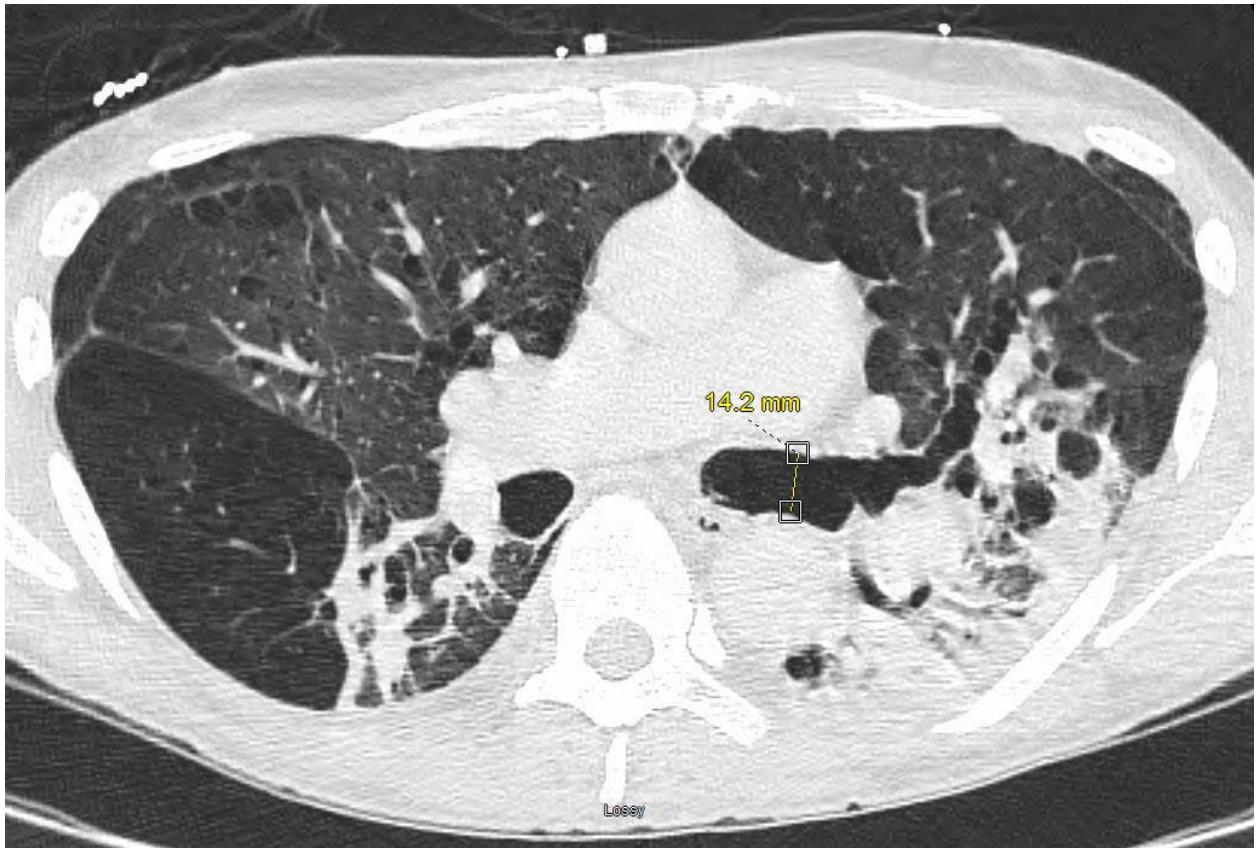
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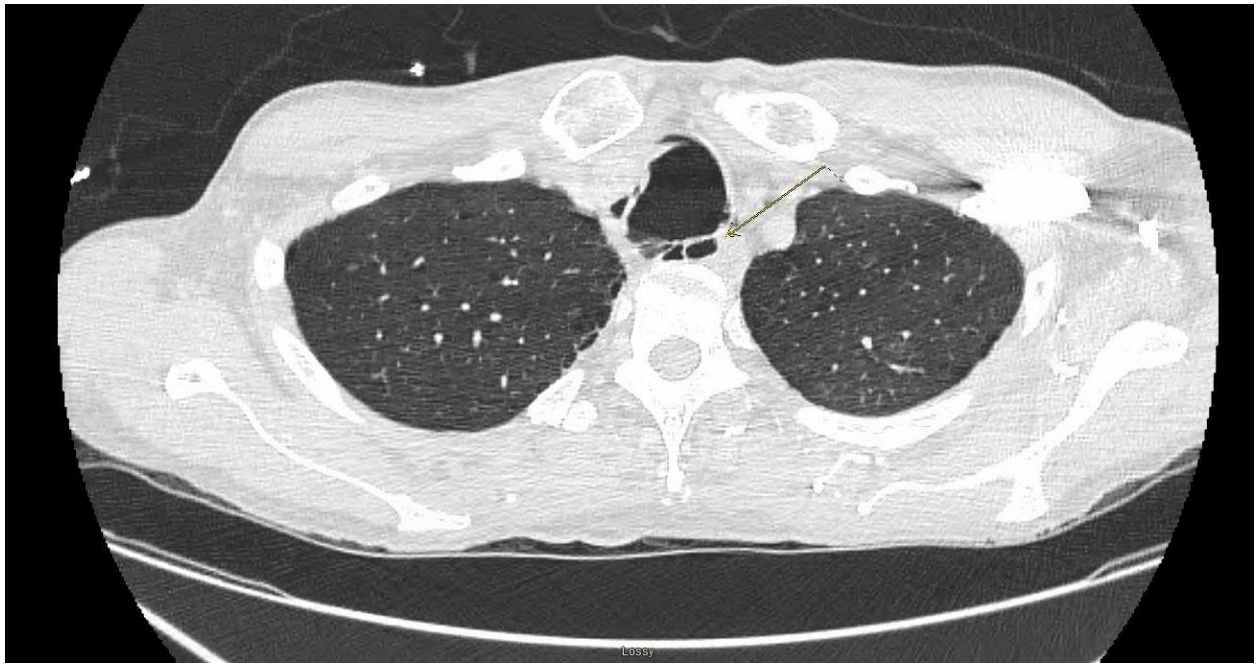

**Figure 1**. Chest computed tomography revealed dilatation of right main bronchus (19.6mm) and bronchiectasis.

## CONCLUSIONS

MKS is associated with various congenital disorders and can present with chronic cough, frequent purulent infection of lower airways, or can be asymptomatic. If a patient with this disorder requires intubation one must consider appropriate ETT size and ventilator settings consistent with those of patients with severe emphysema. One should also anticipate aspiration risk and may even consider empiric antibiotics for ventilator-associated pneumonia.

## REFERENCES

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