

# A Rare Case of Tracheobronchomegaly: The Mounier – Kuhn Syndrome

Kilaru D\*, Stadler C and Alavardian A

Nassau University Medical Center, East Meadow, New York, USA

\*Corresponding author: Kilaru D, Internal Medicine Resident, Nassau University Medical Center, East Meadow, New York, USA, Tel: +5165726501; E-mail: dkilaru@numc.edu

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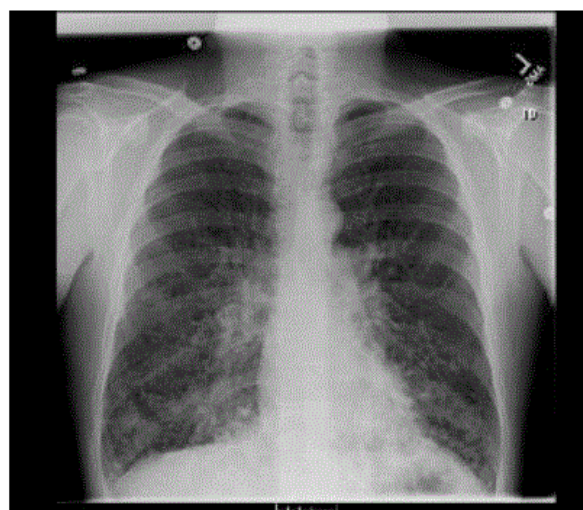
**Keywords:** Pneumonia; Recurrent respiratory tract infections; Cough; Bronchiectasis

## Introduction

Mounier-Kuhn syndrome is a chronic airway disease characterized by dilatation of the trachea and bronchi, recurrent respiratory tract infections and bronchiectasis [1]. It was first described by Mounier-Kuhn in 1932 with fewer than 100 cases reported in the medical literature [2]. Here we present one such case of Mounier-Kuhn syndrome.

## Case Presentation

A 36-year-old Caucasian male presented to the emergency department with shortness of breath, fevers, pleuritic chest pain and productive cough for 10 days. He was treated for pneumonia with four days of azithromycin prior to admission. Patient had a 30-pack-year smoking history and worked in construction. Vitals were BP 121/86, HR 88, RR 18, SpO<sub>2</sub> 86% on room air with diffuse bilateral wheezing and rhonchi. The laboratory findings included a WBC count of 16.32 K/mm<sup>3</sup> with neutrophilic predominance and a bicarbonate of 36 mmol/L. HIV, alpha-1-anti-trypsin, Cystic fibrosis screening mutations, and aspergillus antibody testing were all negative. Chest x-ray revealed persistent basilar predominant thickened and prominent interstitial pulmonary markings, suggestive of chronic lung disease. More patchy/nodular opacities at both lung bases, especially on the left, may represent developing lower lobe pneumonia (Figure 1).



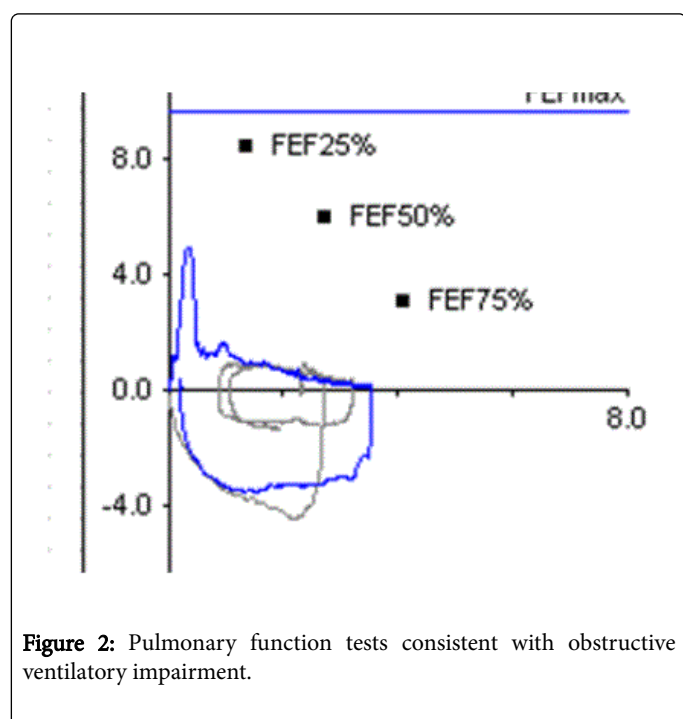
**Figure 1:** Chest x-ray of developing lower lobe pneumonia.

Patient's prior pulmonary function tests were consistent with obstructive ventilatory impairment (Table 1 and Figure 2).

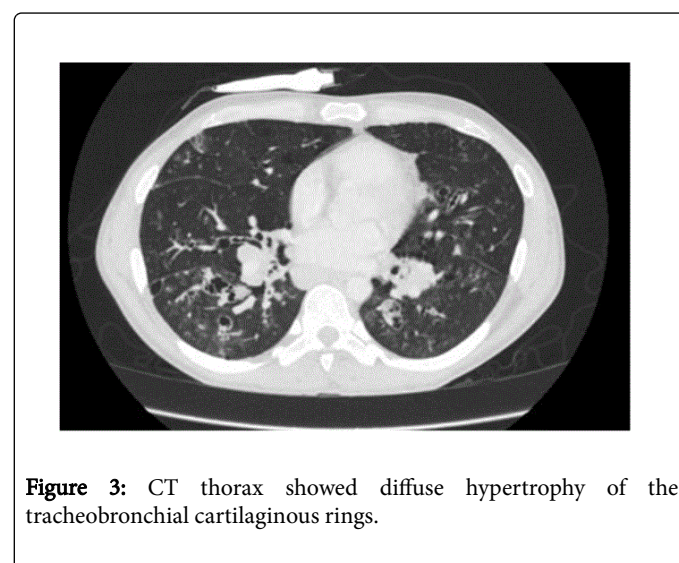
Lung Mechanics	Pre-Bronch				Post-Bronch		
	Units	Actual	Pred	%Pred	Actual	%Pred	%Change
FVC	L,bpts	3.56	5.46	65	-	-	-
FEV1	L,bpts	1.6	4.2	38	-	-	-
FEV1/FVC	%	44.92	76.98	58	-	-	-
FEF25%	L/s	1.41	8.48	17	-	-	-
FEF50%	L/s	0.83	6.02	14	-	-	-
FEF75%	L/s	0.35	3.13	11	-	-	-
FEF MAX	L/s	4.92	9.67	51	-	-	-
FEF25%-75%	L/s	0.67	4.33	15	-	-	-
FIVC	L,bpts	3.34	-	-	-	-	-
FIF50%	L/s	3.36	-	-	-	-	-

FEF50/FIF50	%	24.61	-	-	-	-	-
MVV	L/min,bpts	61.71	150.16	41	-	-	-
P <sub>lmax</sub> /MIP	cmH <sub>2</sub> O	-	-124.3	-	-	-	-
PE <sub>MAX</sub> /MEP	cmH <sub>2</sub> O	-	232.98	-	-	-	-
Lung volumes	Units	Actual	Pred	%Pred	-	-	-
VC	L,bpts	3.62	5.46	66	-	-	-
IC	L,bpts	3.17	3.37	94	-	-	-
ERV	L,bpts	0.45	2.09	22	-	-	-
TV	L,bpts	1.47	-	-	-	-	-
FRC	L,bpts	6.06	4.11	146	-	-	-
RV	L,bpts	5.61	2.02	278	-	-	-
TLC	L,bpts	9.23	7.48	123	-	-	-
RV/TLC	%	60.75	26.98	225	-	-	-
RAW	cmH <sub>2</sub> O/L/s	-	0.20-2.50	-	-	-	-
Sgaw	L/s/cmH <sub>2</sub> O	-	0.11-0.40	-	-	-	-
Diffusion Capacity	Units	Actual	Pred	%Pred	-	-	-
Dsb	ml/min/mmHg	27.63	31.72	87	-	-	-
Vasb	L,bpts	6.74	7.48	90	-	-	-
D/Vasb	ml/min/mmHg/L,bpts	4.1	4.24	97	-	-	-

Table 1: Pre-bronch and post-bronch values.

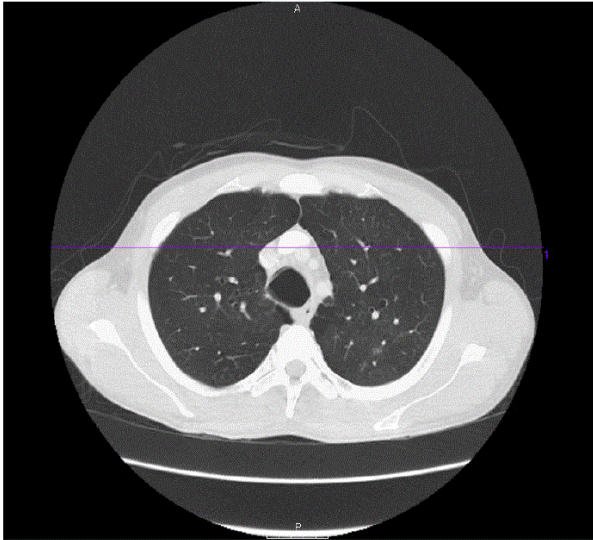


CT thorax showed diffuse hypertrophy of the tracheobronchial cartilaginous rings with tracheobronchial diverticulosis and extensive bilateral bronchiectasis (Figure 3).

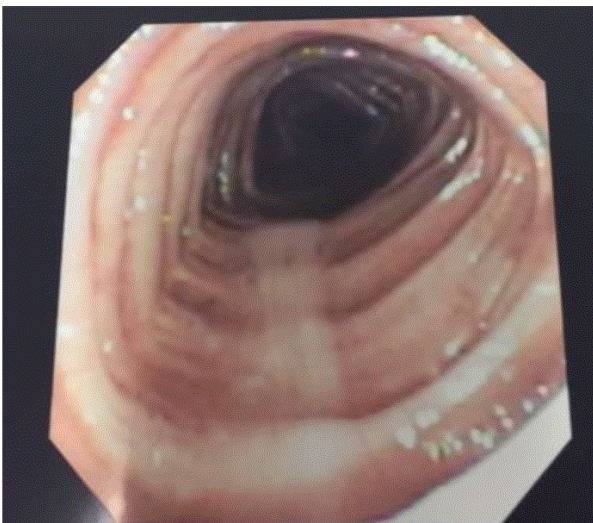


Tracheal diameter was measured to be 33.6 mm (Figure 4), the right and left mainstem bronchi were 32.1 mm and 30.6 mm, respectively. This confirmed the diagnosis of Mounier-Kuhn syndrome. The patient

underwent bronchoscopy that showed an enlarged trachea and mainstem bronchi, with prominent cartilaginous rings (Figure 5).



**Figure 4:** Tracheal diameter of the right and left mainstem bronchi.



**Figure 5:** Bronchoscopy of an enlarged trachea and mainstem bronchi, with prominent cartilaginous rings.

Respiratory culture grew alpha haemolytic streptococcus. The patient was treated for community acquired pneumonia and COPD exacerbation and was discharged home.

## Discussion

Mounier-Kuhn syndrome is characterized by atrophy of longitudinal elastic fibers with thinning of the muscularis mucosa that results in dilation of the membranous and cartilaginous portions of the trachea and main bronchi [1]. Three subtypes exist. In Type 1, there is slight symmetric dilation in the trachea and main bronchi. In Type 2, the dilatation and diverticula are more distinct; and in Type 3 the diverticula extend to the more distal bronchi [2]. Due to this dilatation, patients have an ineffective cough and impaired mucociliary clearance leading to recurrent infections. In adults the diagnosis is made on CT scan if the trachea is  $>30$  mm, the right main bronchus is  $>20$  mm, and the left main bronchus is  $>18$  mm [2]. Treatment is supportive and includes mucolytic treatment and postural drainage to counteract the impaired mucociliary clearance [1]. Patients should receive pneumococcal and yearly influenza vaccines regardless of age and be counseled in smoking cessation [1,3]. Because of easily collapsible airways, tracheobronchoplasty and airway stenting have been performed. Lung transplant has also been reported twice in patients with Mounier Kuhn syndrome [1].

## Conclusion

This case presentation is an example of a patient that was misdiagnosed as a case of COPD rather than Mounier-Kuhn syndrome. This was due to a confounding variable of smoking, however, the patient's young age and enlarged airways on CT thorax revealed his diagnosis of Mounier-Kuhn syndrome which also explained his recurrent pneumonias.

## References

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