



# Mounier Kuhn bronchoscopic and radiological findings.

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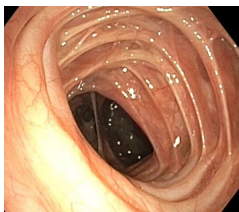
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## Introduction:

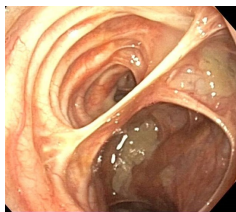
- Mounier-Kuhn syndrome, is a rare clinical and radiologic condition characterized by marked tracheobronchial dilation and recurrent lower respiratory tract infections.
- It is characterized by tracheobronchial dilation secondary to atrophy of the muscular and elastic tissues in the trachea and main bronchial walls.
- Diagnosis is established with computed tomography (CT) confirmed by bronchoscopy, as well as pulmonary function testing.

## Case presentation:

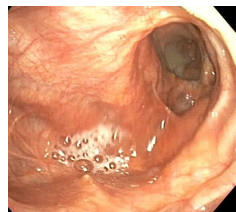
- A 32 years old female patient followed up for a Mounier Kuhn syndrome. She was referred to our center for lung transplantation evaluation.
- CT scan showed tracheobronchomegaly, 35 mm tracheal diameter and 28 mm for the left main bronchus (Fig: 1,2), multiple tracheal diverticula mainly in the posterior tracheal wall, varicose and cystic bronchiectasis extended to the distal bronchi.
- Fiberoptic bronchoscopy was consistent with the CT findings.
- Pulmonary function test: FVC 63% 2180mL, FEV1 42% 1270mL, FEV1/FVC 58%, TLC 99% 4840mL, RV 191% 2760mL, DLCO 48%, KCO 68%.



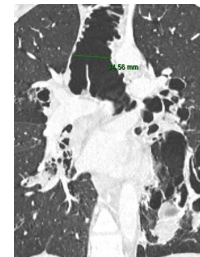
Trachea



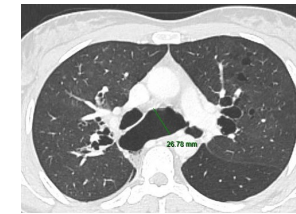
Carina



LMB



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## Conclusion:

- Mounier-Kuhn syndrome is secondary to a thinning of the muscular mucosa and atrophy of the longitudinal muscle and elastic fibers of the tracheobronchial tree.
- As a consequence, tracheal diverticulosis and dilations in the posterior membranous wall appear, along with bronchiectasis that tend to be cystic in appearance.
- Overall, there is an impairment of mucociliary clearance, with an ineffective cough, which predisposes the patient to recurrent lower respiratory tract infections.
- Three subtypes were described in literature: type 1 with subtle symmetrical dilation of trachea and major bronchi; type 2 with more obvious dilation and eccentric diverticula; type 3 with diverticula that extend to the distal bronchi.
- Treatment ranges from respiratory physiotherapy for airway clearance to endobronchial stenting in severe cases.
- Lung transplantation is not technically possible due to absence of bronchial airway required for the anastomosis.