

# Mounier Kuhn Syndrome

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#### Declaration of conflicting interests

The authors declare that there is no conflict of interest.

#### Abstract

**Background:** Tracheobronchomegaly or Mounier Kuhn Syndrome is a rare disorder of uncertain etiology characterized by marked dilatation of trachea and bronchia and recurrent respiratory infections. We report a case of 19 years old female who presented with recurrent respiratory tract infections.

**Key Words:** Mounier-Kuhn Syndrome; TBM; Pakistan

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## Introduction

**M**ounier-Kuhn Syndrome or TBM is a rare clinical and radiological entity characterized by marked dilatation of trachea and bronchi and recurrent Respiratory tract infections.<sup>1-3</sup>

MKS is also known by different names like tracheiecteris, tracheomegaly, Tracheobronchopathia malacia, multiple tracheal diverticulae.

## Case Report

A 19 years old female patient was admitted with complaints of recurrent lower respiratory tract infections since 05 years of her age, presented with episodes of productive cough and fever for last 03 months, there was also history of grade 01 dyspnea, however no hemoptysis, orthopnea and paroxysmal nocturnal dyspnea. Frequency of Lower respiratory tract infections have increased for last 04 years. GPE was unremarkable other than slightly diminished air entry at lung bases.

Labs investigations revealed normal leukocyte count, renal parameters and serum electrolytes. Chest X-ray showed heterogenous opacity in left lower radiological zone extending from hilum to dome of diaphragm e internal air bronchograms.

CT Chest was performed and revealed bronchieactatic changes involving posterior segments of RUL Ct Chest e MPR (multi planar

reconstruction) was obtained that showed dilatation of trachea measuring 29.9 mm, 02 cm above the level of aortic arch, dilatation of right main bronchus and left main stem bronchus measuring 26.3 and 29.9 mm respectively. CT also clearly showed cystic bronchiectasis of Right upper lobe and left lower lobe.

Fiberoptic bronchoscopy revealed dilated trachea and widening of bronchial tree bilaterally with flaccid walls and diverticula outpouching in main bronchi.

## Discussion

Mounier-Kuhn syndrome in 1932 described a rare entity characterized by dilatation of trachea and bronchi with absence of elastic fibers and thinning of smooth muscles in the 3rd or 4th decade of life with recurrent respiratory tract infections.<sup>5</sup>

Incidence of TBM is rare, however actual number of cases may b greater as some pts are asymptomatic and those who are symptomatic, TBM is frequently overlooked when chest Radiograph are made.<sup>6</sup> The other conditions such as Ehlers Danlos Syndrome, Marfan syndrome and Lutis Laxa may also present with secondary tracheobronchial dilatation.<sup>7</sup> however most cases are sporadic and idiopathic and shows no evidence of associated connective tissue disorder.<sup>8</sup>

On CT scan the diagnosis is made when the transverse diameter of trachea measures greater than 03 cm and that of right and left bronchi exceeds 2.4 cm and 2.3

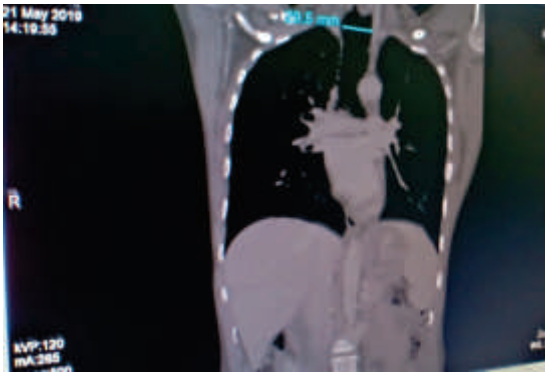


Fig 1: CT Thorax with MPR shows abnormal dilatation of trachea 2.95cm.

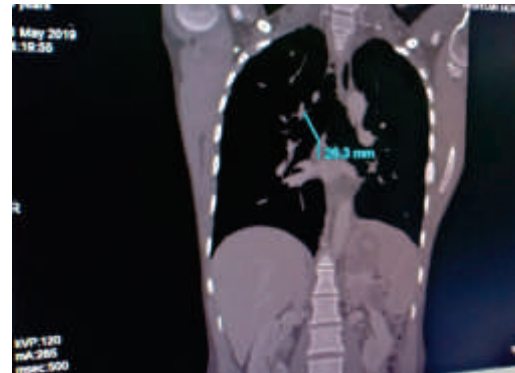
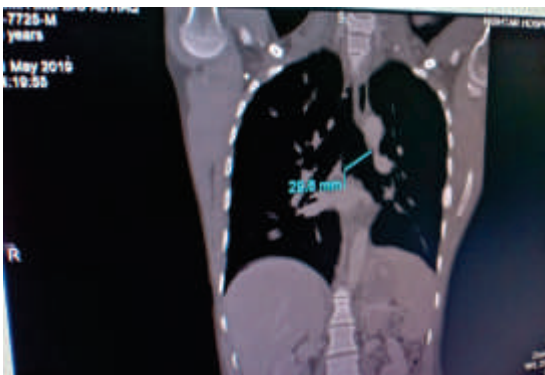


Fig 2: CT thorax with MPR shows right mainstem bronchus diameter as 2.63cm



CT thorax with MPR shows left mainstem bronchus diameter as 2.96 cm.



Fig 4: CT thorax with MPR shows abnormally dilated trachea and mainstem bronchi.

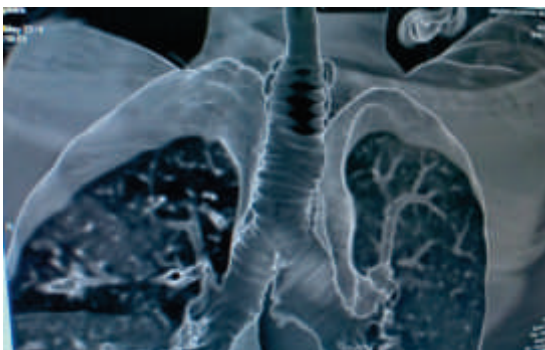


Fig 5: CT thorax with MPR shows abnormally dilated trachea and mainstem bronchi

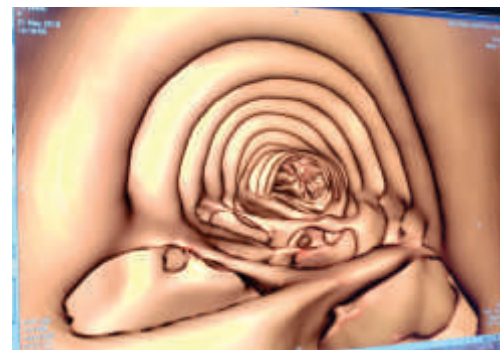


Fig 6: CT thorax with MPR shows abnormally dilated trachea

cm respectively. The diameters in our case were 2.95, 2.6 and 2.9 cm respectively,9 furthermore in our case CT also showed bronchieactasis and bronchoscopy showed bronchial diverticulosis which confirmed the diagnosis.

In symptomatic patients chest physiotherapy, postural drainage and antibiotics are mainstay of treatment for clearing secretions and suppressing infections.

Surgery is rarely done due to diffuse nature of disease

with tracheal stenting being useful in advanced cases with report of tracheobroncheal endoprosthesis being used with success.<sup>10-12</sup>

As tracheobronchomegaly are often overlooked on plain chest x rays, patients with chronic and recurrent respiratory tract infections should have CT scan done to rule out this condition.

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