

# CASE REPORT

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## TRACHEOBRONCHOMEGALY: MOUNIER-KUHN SYNDROME: A CASE REPORT WITH REVIEW OF LITERATURE

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**INTRODUCTION:** Mounier-Kuhn syndrome is a rare clinical and radiological entity characterized by marked dilatation of trachea, bronchi, bronchiectasis and recurrent lower respiratory infections. Etiology of this disorder is uncertain and clinical presentation variable.

**CASE REPORT:** An 85 years old female patient presented with worsening cough with expectoration, scanty, white, mucoid, non-foul smelling, non-blood tinged, increases at night; breathlessness, insidious in onset, gradually progressed, increases on exertion, decreases at rest and fever since 15 days. Patient gave history of bilateral vague chest pain and difficulty in expectorating sputum. Previous history of similar recurrent episodes of LRTI was present since childhood.

On examination: patient was having ineffective cough. Inspection, palpation and percussion were unremarkable. On auscultation, bilateral coarse crepitations were heard mainly in the infrascapular region. Other system examination was normal.

On evaluation: Hematology: (Hb-12.1 g/dl, WBC count-10, 320 cells/cumm, peripheral smear- normocytic normochromic blood picture), Renal function test and Liver functions were within normal limits. Sputum for AFB – Negative and sputum culture yielded commensals.

Chest-X-Ray showed multiple air cysts of varying size in bilateral lower zones suggestive of bronchiectasis with enlarged tracheobronchial air shadow.

CT scan Thorax revealed- tracheobronchomegaly with tracheal diameter to be 30mm and right main bronchus and left main bronchus 24.1 mm and 20.4 mm respectively with sacculations in between the tracheal rings.

Pulmonary function tests – showed obstruction of both large and small airways with poor reversibility

Patient refused bronchoscopy and bronchial biopsy.

**Treatment:** Exacerbation of bronchiectasis was managed conservatively with antibiotics, mucolytics and postural drainage. At discharge, patient was put on LABA and low dose ICS. Patient is being followed up regularly and is symptomatically better without any exacerbations.

**DISCUSSION:** Mounier-Kuhn syndrome or tracheobronchomegaly, is a rare clinical and radiological entity described for the first time by Mounier and Kuhn in 1932. The syndrome is characterised by marked tracheobronchial dilatation with recurrent respiratory tract infections.<sup>1</sup> Fewer than 100 cases have been reported in medical literature.<sup>2</sup> Satish Kachhawa et al reported a

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case of Mounier-Kuhn syndrome from Bikaner, India.<sup>3</sup> P Jain et al reported a case of variant of Munier-Kuhn syndrome from Udaipur.<sup>4</sup>

Although etiology is uncertain it is believed to be due to the atrophy or absence of elastic connective tissue and thinning of smooth muscle layer in the trachea and main bronchi (when examined histo chemically with Verhoeff elastic stain), leading to sacculations and the formation of diverticula between the cartilage rings.<sup>5, 6</sup> It also might be due to absence of myenteric plexus of bronchial tree.<sup>7</sup>

The airways are thus flaccid and markedly dilated on inspiration and collapsed on expiration or cough. The abnormal airways dynamics and pooling of secretions in broad out pouching's of redundant musculomembranous tissue between the cartilaginous rings and there is ineffective cough with impaired mucociliary clearance that predisposes to development of chronic pulmonary suppuration, bronchiectasis leading to emphysema and non-specific pulmonary fibrosis.<sup>8</sup>

Mounier-Kuhn syndrome has 3 subtypes. In type 1, there slight symmetrical dilatation of trachea and main bronchi. In type 2, dilation and diverticula are distinct. In type 3, diverticular and saccular structures extend to distal bronchi.<sup>9</sup>

Disorders such as sarcoidosis, usual interstitial pneumonia and cystic fibrosis, which cause severe fibrosis of the upper lobes, may also exert sufficient tracheal traction to result in tracheal enlargement. Certain other conditions such as Marfan syndrome, Ehlers-Danlos syndrome, Kenny Caffey syndrome, Ataxia telangiectasia, connective tissue diseases, Brachmann-de Lange syndrome, Bruton-type agammaglobulinaemia, Ankylosing spondylitis, Cutis laxa, and light chain deposition disease are also associated with secondary tracheobronchial enlargement. Most cases are sporadic and show no evidence of associated connective tissue disease, as was the case in our patient also.<sup>10</sup>

On X-ray chest, enlarged tracheobronchial air shadow is seen often with bronchiectasis. Two phase radiographs will demonstrate enlargement of trachea on inspiration and collapse during expiration, but less sensitive.<sup>11</sup>

CT scan Thorax will reveal trachebronchomegaly. Multiple diverticula and areas of scalloping to be seen between the cartilaginous rings in the trachea and both main bronchi with cystic bronchiectasis bilaterally.

		Female	Male	Our patient
Trachea	Transverse	>21mm	>25mm	28mm
	Sagittal	>23mm	>27mm	30mm
Right main bronchus		>19.8mm	>21.1mm	24.1mm
Left main bronchus		>17.4mm	>18.4mm	20.4mm

Upon pulmonary function testing, decreased bronchial flow speed, increased tidal volume and dead spaces and obstructive pattern may be observed.<sup>12</sup> Other tests to be considered are DLCO, alpha 1 antitrypsin titre hypersensitivity precipitin testing and ABG if required.

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Fibreoptic bronchoscopy will have features of dilated trachea with prominent tracheal rings and widening of the bronchial tree bilaterally with mucopurulent secretions. Expiration test reveals tracheobronchial collapse.

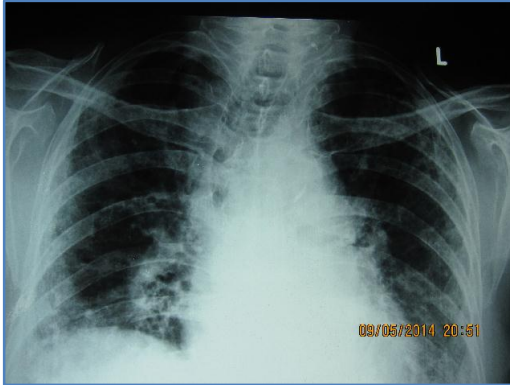
Management of symptomatic patients would consist of intensive and appropriate antibiotic therapy, general supportive therapy e.g: bronchodilators and postural drainage. Bronchoscopy, tracheostomy or both may facilitate the management of secretions and mucus plugging in some patients. In few reported cases Y shaped tracheobronchial stent placement has helped the trachea to remain open with significant clinical improvement.<sup>13</sup>

**CONCLUSION:** Diagnosis of Mounier-Kuhn syndrome, though rare, can be considered in patients with recurrent LRTI with bronchiectasis. Diagnosis can be made easily with chest-X-ray and CT scan thorax.

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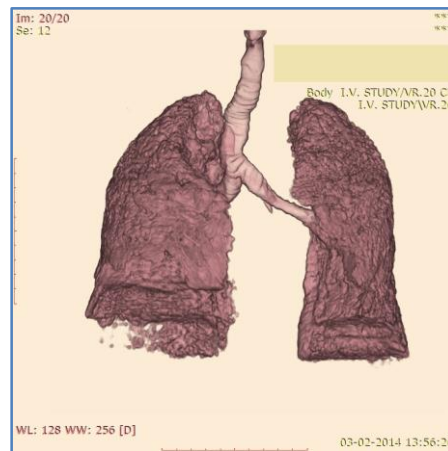
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**Fig. 1**



**Fig. 2**



**Fig. 3**

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