Respiratory Medicine (2013) 107, 1822–1828

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REVIEW

Mounier-Kuhn syndrome or congenital tracheobronchomegaly: A literature review

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Received 30 April 2013; accepted 29 August 2013
Available online 6 September 2013

KEYWORDS
Mounier-Kuhn syndrome;
Tracheo-bronchomegaly;
Chronic respiratory infections;
COPD;
Bronchiectasis

Summary
Mounier-Kuhn syndrome or congenital tracheobronchomegaly is a chronic airway condition which for currently unknown reasons mostly affects males. It is commonly overlooked on conventional chest X-rays, and is considered to be rare, but the prevalence might be higher as commonly assumed. The hallmark of it is a dilatation of the main airways which frequently, but not always, causes marked, mainly respiratory, symptoms, and patients usually present with varying degrees of recurrent infections, breathlessness, haemoptysis, dyspnoea. Although at least 200 case reports have been published, there have been only a few attempts to review them, and none in the last 20 years. Due to the lack of clinical trials and wide variability of case-report format, a systematic review was deemed not feasible, therefore PubMed and Medline databases were searched using terms "Mounier-Kuhn syndrome", "tracheobronchomegaly", "tracheomegaly", and "bronchomegaly", without any time restrictions, to summarize currently known facts about the syndrome. To the authors' best knowledge, the result is currently the most comprehensive review of previously published literature about the congenital tracheobronchomegaly, and summarizes what's known about symptoms, prevalence, disease associations, and treatment options for this syndrome. TBM — tracheobronchomegaly, MKS — Mounier-Kuhn syndrome, CT — computed tomography, COPD — chronic obstructive pulmonary disease.

Abbreviations: TBM, tracheobronchomegaly; MKS, Mounier-Kuhn syndrome; CT, computed tomography; COPD, chronic obstructive pulmonary disease.

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http://dx.doi.org/10.1016/j.rmed.2013.08.042

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Introduction

The histological findings of enlarged airways were described already in 1897 [S1] but only in 1932 the first clinical description was made [S2]. Many similar cases of trachea- and bronchomegaly have been subsequently published, both as isolated cases and as cases where it goes together with some other condition. With greater recognition of the syndrome, it is the latter type of reports that are becoming increasingly more common. But, despite the growing number of cases published, reviews of them are few. The first comprehensive review articles were first published in 1962 (by then the eponym Mounier-Kuhn syndrome (MKS) was already in use) [1] and in 1965 [2]. Some thirty years later a report of a series of ten cases again reviewed the most important characteristics of the syndrome [3], but none of authors, since Katz [1] & Johnston [2], have attempted to create a comprehensive review of available literature on the MKS. Most of them cite just a handful of articles published up to now. More importantly, the number of case reports published since the last review has increased significantly, including a variety of new treatment options for these patients. The first descriptions of long-term follow-up start to emerge and at least one clinical trial has been conducted since the last review in 1991. Due to the lack of clinical trials or epidemiological studies and wide variability of published case reports, we lacked an appropriate basis for a systematic review and therefore the following pages summarize the current knowledge about the MKS as can be concluded from published cases — by presenting both the available facts and contradictions about the pathogenesis, symptoms and possible management of the disease, showing the wide variability of the MKS and different problems caused by it.

Methods of literature search and description of results

Both EK and ZK independently searched the PubMed and Medline databases for the terms "Mounier Kuhn syndrome", "Tracheobronchomegaly", "Tracheomegaly" or "Bronchomegaly". No publication date restrictions were used. Abstracts were then reviewed to identify any relevant publication and full text was then retrieved if it was estimated that it would provide valuable additional information. If opinions of EK and ZK differed the opinion of AB was deciding. A similar search was made in Google Scholar and

Table 1 Airway dimensions of adults, normal values, mm.

	Gender	Subject age	Number of subjects	Trachea		Left main bronchus	Right main bronchus
				Sagittal section	Coronal section		
Himalstein & Gallagher [S24]	Males	29—91 (mean 65, median 71)	100	24.7	28.6	22.5	23.8
Bretnach [7]	Males	10—79 (mean & median n/a)[a]	430	27	25	—	—
	Females		378	23	21	—	—
Roditi [8]	Males	18—82 (mean 56)[a]	47	27.4	24.9	—	—
	Females		28	21	20	—	—
Woodring [3]	Males	16—79 (mean & median n/a)[a]	79	—	—	18.4	21.1
	Females		121	—	—	17.4	19.8

[a] Age for males/females separately not given in the publication.