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REVIEW

Mounier-Kuhn syndrome or congenital tracheobronchomegaly: A literature review

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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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Contents

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.

references of the acquired articles were reviewed to identify any additional publication. If the language of an article wasn't English, German or Russian, possibility of translation was evaluated. Publications not-indexed in PubMed or Medline were considered if in authors' opinion they provided relevant information to this review.

A search for MKS in PubMed returned over 260 results and most were about the syndrome in question. Published articles ranged from reports of single patients with typical symptoms and/or associations with other diseases to a few articles presenting a series of up to ten patients [3—5]. A single clinical trial has been conducted [5]. Only a few authors have followed patients for more than a year [S3,S4], most authors are describing patients at the time when their diagnosis was made. Majority of articles are published in English but there are reports available in many languages [6], [S5—S19].

Review

Defining normal airways

The first review defined the MKS as "a distinctive clinical—roentgenologic condition consisting of marked dilatation of the trachea and major bronchi and associated with chronic respiratory tract infections" [1]. But to understand what is enlarged or dilated trachea one first has to agree upon what is the size of normal airways and several studies have attempted that.

Table 1 shows studies on normal airway values in adults (discussing details about the children is beyond the scope of this paper and the interested reader is referred to the works of Griscom [S20—S22] or de Jong [S23]). Most importantly, in 1984 [7], using the conventional chest X-rays, it was found that the size of the trachea was dependent only from the gender, and not from the body mass or height. The author, assuming that the size of a human trachea follows a normal distribution, suggested that the upper limit should be two standard deviations above the average values, when measured 2 cm above the projected top of the aortic arch (although no significant differences were noted in measurements on other levels) [7]. No bronchial measurements were done in this study. These were added later [3] (Table 1).

A study of 150 patients in 1994 confirmed that the same values can be used to evaluate measurements done by CT [8].

General characteristics in MKS

The typical histological findings of MKS were summarized by Katz already in 1962 [1]. The thinning of muscular mucosa, as well as the atrophy of longitudinal muscle and elastic fibres are universal and accepted hallmarks of MKS [1], [S25]. Numerous saccular diverticula between cartilages, and bulging and spindle shaped dilatations in the posterior wall may be seen. It's assumed that these arise when redundant non-muscular segments protrude between the transverse bands of muscle that connects the tips of U shaped tracheal cartilages [1]. The absence of myenteric plexus in the tracheal wall may suggest a condition similar to Hirschprung's disease or achalasia of gastric cardia [S26].

A suggestion to classify all cases of TBM in three types based on anatomical appearance has been made [S24]. Type I would have symmetrical diffuse enlargement of both trachea and bronchi. In the most common — Type II, the enlargement is more eccentric with pronounced diverticula and abrupt change to normal bronchial size. Lastly, the Type III, where diverticula may extend to the more distal bronchi but only few case-reports have used it.

To the best knowledge of the authors no population based studies of MKS have been published so far. But some publications estimate a prevalence between 0.4 and 1.6% (Table 2) in patients with pulmonary symptoms.

Nevertheless the 7 cases out of 10 reported by Woodring [3] that were found accidentally, and the fact that enlarged trachea is easily overlooked in chest X-ray [S31] casts doubt whether the syndrome is really so rare. The number of published case reports grows steadily — a possible sign of greater awareness. Additionally, intentional searching for cases would identify minimal tracheal enlargement that would otherwise remain overlooked [8].

MKS may be mistaken with Williams-Campbell syndrome [S32], a congenital deficiency of the 4th to the 6th order bronchial cartilages [S33]. But on CT scans the bronchial defects of the Williams-Campbell syndrome begin at the bronchi of the 4th order division — a level where the enlarged airways of MKS should return to normal and in Williams-Campbell syndrome the collapse of cystic bronchial dilatations can be observed in expiratory CT scans [S34,S35].

The age of patients with MKS varies greatly: from a 18 months old girl [S36] and a 19 months old boy [S37] to an 86 years old [S38] man (a case of a 15 months old boy [S39] with involvement of the bronchi of sixth and seventh division can't be clearly attributed to MKS [9]). For most

Table 2 Prevalence studies for TBM.

Author	Year	Prevalence	% of patients
Fiser [S27]	1969	6 cases in 1200 bronchograms	0.5%
Abramovic [S28]	1970	3 cases in 500 bronchograms	0.6%
Himalstein [S24]	1973	5 cases in 500 bronchograms	1%
Bateson [S26]	1973	2 cases in 120 bronchograms	1.6%
Acimovic [S29]	1994	4 cases in 396 bronchograms	1.01%
Roditi [8]	1994	7/42 CTs of patients with bronchiectasis	16.6%
Cartier [S30]	1998	1/82 CTs of patients with bronchiectasis	1.2%
Menon [4]	2008	8/5234 pulmonary CTs (of symptomatic patients)	0.15%

patients the diagnosis is made only after the third decade of life; In one report [2] the average age of patients was 39 years, and in two others [3,4] — 56 years. But it's not uncommon that patients are diagnosed only in their 70-ties [S40], despite a long history of respiratory symptoms, as shown by the case of the 79 years old woman, who was diagnosed with chronic obstructive pulmonary disease (COPD) despite being life-long non-smoker and with no relevant occupational history [S41].

There is a strong male predominance of about 8:1 [2,4] and most of the patients seem to be smokers, although life-time non-smokers have been reported [3], [S41,S42]. A possibly accidental predominance in Afro-American patients has been noted [S26].

The origin of MKS seems to be congenital, as confirmed by the histo-pathological data, in association with recurrent childhood infections and occasional occurrence of MKS together with several congenital disorders. A description of two siblings with MKS [2] and a possible case of MKS among cousins [3] has caused some to think of autosomal recessive disorder [2].

Although the previously cited definition does not mention the possible aetiology, the authors of this review hold the opinion that only cases without any clear cause should be labelled as MKS and patient's history should be carefully taken and reviewed if TBM is suspected as a list of conditions may cause acquired TBM: pulmonary fibrosis [10], [S43], mechanical ventilation [S44] especially for preterm neonates [S45] and radiotherapy [S46]. TBM due to the pulmonary fibrosis may be less marked as in cases without a clear cause [10]. Additionally several immunodeficiency syndromes in children [S47] have been associated with TBM. Neonatal TBM is a common complication following intrauterine tracheal occlusion [S48,S49]. A case secondary to tuberculosis has been published, but the airway size was not given [S50].

Table 3 List of diseases reported together with MKS.

Adrenal insufficiency [S63]
Amyloidosis, Type AA, Secondary [S13]
End-stage renal disease (due to arterial hypertension with renal transplantation) [S3]
Fibrous histiocytoma [S64] [a]
HIV [S10]
IgA nephropathy [S65]
Isolated adrenocorticotrophic-hormone deficiency [S66]
Laryngeal cancer [S67]
Light chain deposition disease [S17]
Non-small cell carcinoma [S68]
Non-tuberculous mycobacterial infection [S69]
Oesophageal carcinoma [S70]
Pleomorphic carcinoma [S18,S71]
Ovarian cancer [S72]
Renal tumour [S65] [b]
Tracheopathia osteoplastica [S73]
T-cell lymphoma [S38]

[a] Publication had no data about the airway size.
[b] No further information available.

Associations with other diseases

Several case reports mention congenital defects, including accessory tracheal bronchi [S19,S27,S29,S51], but as for one report [S51] the airway size is not given, the criteria for the syndrome can't be verified. TBM has been described in 3 cases of Brachmann-de Lange [S52], in a case of Kenny-Caffey syndrome [S53] and one case includes abnormal bronchial vasculature [S29].

There are cases of TBM reported together with Ehlers-Danlos [11], [S54] and Marfan's [S55] syndromes and cutis laxa [S56] as well as rheumatologic disorders — ankylosing spondylitis [12], [S42,S57,S58] (although for one study [S58] the criteria of tracheobronchial dilation are not clear) and two patients with rheumatoid arthritis [13,14]. In a child with Ehlers-Danlos syndrome [S59] the airway size didn't quite reach the criteria for MKS but it can't be excluded that the trachea might have enlarged further reaching the necessary criteria of TBM. To complicate things further, a patient with Marfan's syndrome developed a significant tracheomegaly after therapeutic irradiation [S60]. And there is a list of indistinct cases where the accompanying disease of MKS has not been clearly diagnosed [15], [S19,S61,S62].

Table 3 lists the conditions reported together with MKS that most likely should be looked at as separate problems (in alphabetical order).

Clinical findings of MKS

The complaints are non-specific and largely related to the infectious consequences of the disease. In many patients they begin in third decade of life. Chronic, recurrent respiratory infections with productive or dry cough [S40] and purulent sputum, dyspnoea (occasionally on exertion) [S74] and haemoptysis [5], [S16,S29] all seem to be common. Some patients have had recurrent infections since early childhood [6] and others are diagnosed accidentally [16], [S75] having had no symptoms yet, but it seem that only a handful remain symptom-free lifelong. The recurrent respiratory complaints frequently are interpreted as caused by COPD, even when no criteria support it [17], [S41]. In developing countries some have received empiric therapy for tuberculosis [18], occasionally without evidence of the presence of tuberculous bacteria [S76,S77]. Progressive hoarseness, caused by vocal cord paralysis, possibly due to changed crycoarytenoid joint, has been described [S61].

It is unclear whether the TBM is progressive. Only two patients have been observed from childhood to adulthood. One was first diagnosed with asthma at the age of 4. Bronchiectasis were noted at the age of 13, and after four years MKS was diagnosed [S4]. The other was diagnosed at the age of 11 [S3] and the tracheal size didn't change significantly in later years. One adult case with TBM [S78] had no increase in the tracheal size over 8 years, but the patient had pulmonary fibrosis which can cause acquired TBM [10], [S43].

Upon clinical examination, finger clubbing is common, and bronchial rales and/or wheezing can be heard upon auscultation. Fever, chills, tachypnoea and tachycardia may be present during exacerbations. Gastroesophageal reflux disease [5] may be present and the syndrome may

contribute to or at least exacerbate symptoms of the sleep apnoe [S79].

Varying degree of obstruction and an increased residual capacity are noted on spirometry [15] and values are lower during the infectious exacerbations [6] but respiratory function may be normal [S9]. Together with dynamic CT, spirometry can help documenting tracheal collapse [S80].

TBM is visible on plain chest X-rays [3] where the tracheal size may exceed the width of the vertebral column [1], [S54], but it's not uncommon to miss it altogether until a CT is done [17]. An abrupt change from dilated to normal calibre bronchi in peripheral airways has been noted with a suggestion that in acquired bronchomegaly the airways would dilate symmetrically [9]. CT has become the golden standard for confirming the diagnosis since its first use for MKS in 1988 [19], [S81,S82]. Use of magnetic resonance imaging is limited to one case-report, and provided no significant additional information [20].

During bronchoscopy the increased tracheal diameter and the expiratory collapse due to tracheomalacia [S83] is seen and the redundant tracheal wall may even obstruct the view [1]. In an endobronchial confocal fluorescence microscopy a patient with MKS had intense bronchial autofluorescence and decreased general fluorescence intensity, with spectral shape of normal tissue, that, according to the authors of the study, suggests the disappearance of the fibred connective network of the bronchial wall [21].

The variability of the disease is at best represented by both cases of long-term follow-up. The one patient suffered recurrent feverish respiratory infections but retained normal pulmonary function [S3] whereas for the other fever was uncommon but the pulmonary function steadily dwindled reaching severe obstruction [S4].

The bullous emphysema is common in MKS and may cause pneumothorax [11], [S84—S86] in otherwise asymptomatic patients with TBM [S14]. It has been suggested that these cases should be treated alike along the guidelines for secondary pneumothorax in COPD [S84].

Two cases of aspergillosis and MKS are known. In one case, in which aspergillosis was diagnosed after pulmonectomy due to massive bleeding [S16], the MKS was already diagnosed but in the second case TBM and the allergic aspergillosis were diagnosed at the same time [S87].

Surgery and anaesthesia

MKS causes difficulties across many specialities but especially anaesthetist's work is complicated by the large and weak airways, inefficient cough mechanism, tracheal diverticula, endotracheal tube dislodgement and possible postoperative tracheal collapse [16], [S63]. Endotracheal tube cuff-leak during endotracheal intubation might be the first sign of MKS [S70,S88]. A partial oesophagectomy had to be completed with partially deflated lung due to persistent inability to achieve air-seal during the intubation [S70]. A marked tracheal stenosis after intubation of a patient with TBM demanded surgical intervention [16] so the use of uncuffed endotracheal tube was suggested [16] or the laryngeal mask may be used [S72]. An excellent review of possible solutions while providing anaesthesia to MKS patients is given by Kim et al. [S44]. But not all cases of TBM have operative complications [S3,S65,S75].

A study, including some patients with MKS, showed that surgical tracheobronchoplasty may be used to correct tracheomalacia [22]. And several patients with TBM have had other surgical interventions with varying degrees of success: uneventful renal transplantation [S3], nephrectomy [S65], lower left pulmonary lobectomy [S89] and pulmonectomy [S16], an elective repair of epigastric hernia [S90], septoplasty [S63], an emergency off-pump coronary bypass [S75], and bilateral resection of pulmonary bullae via median sternotomy approach [S91] all have been reported. One patient with TBM developed hernia of the lung through thoracic wall, after resection of a non-small cell carcinoma [S68].

Treatment options

Mucolytic treatment and physical therapy, including massage and postural drainage, have been used to counteract the decreased clearing ability of bronchial tree, to increase the separation of the sputum and to facilitate expectoration. It has been noted that a pneumococcal polysaccharide vaccine provides at least a temporary relief [S4] and vaccination against influenza has been previously suggested [23].

Trachebronchomalacy is present when more than 50% of the airway collapse during expiration [24] and alone [S92] or when complicated by stenosis [S83] it may cause serious breathing problems. Non-invasive continuous positive airway pressure ventilation has shown promising results in reducing the symptoms and treatment was successful in several cases of TBM [18], [S83,S92]. About 8—10 mmHg positive airway pressure is usually sufficient [18], [S92].

Airway stenting and/or tracheobronchoplasty for tracheomalacy is supported by several trials [24,25] which have included patients with MKS. One trial included 3 patients with TBM and showed that silicone stents improve respiratory symptoms and quality of life in patients with tracheobronchomalacy [25]. A single patient was included in a report of advantages of tracheal T-tube which suggested that tracheal T-tube ought to be the preferred management for chronic airway obstruction, if it cannot be corrected by surgery [S93]. One clinical trial (with 12 patients) showed [5] that an aggressive approach with airway stenting and surgical tracheobronchoplasty achieves better results than supportive measures alone [5].

Good results with tracheal stenting are seen in single case reports. Y shaped silicone stent achieved full rehabilitation in social and professional life for a patient with both TBM and tracheal stenosis [S83]. Marked improvement was achieved with a self-expanding metal [S7] and a Dynamic [S94] stent. In the latter case the result persisted at least for a year. Symptoms improved in another case but the type of stent and the extent of improvement were not given [S5]. Availability of appropriate airway stent may be a significant problem as the diameter of most commercial stents does not exceed 20 mm [S55], although these have been used with good results in exceedingly large (up to 42 mm) airways. A custom made stent may be used [S55].

In one case the posterior wall collapse has been successfully treated with Endoscopical laser treatment. Four procedures over 8 years achieved good results [26]. Laser devascularizes and coagulates the tissue causing a retraction of submucousa which rigidifies the posterior membrane of trachea [26].

Lung transplantation has been reported twice: a bilateral lung transplantation in a patient with 27 years of pulmonary symptoms and rheumatoid arthritis [14] and in a patient, whom the MKS was diagnosed in the explanted lungs [27]. Both patients died soon after the transplantation [28]. It is interesting that Ng [S88] mentions another patient with mild TBM who had received a double lung transplant. Unfortunately no additional information is given, but if this is true it seems to be the only and unreported successful lung transplantation in a MKS patient.

Discussion and conclusions

At least two decades have passed since the last significant review of the available literature on MKS, published in 1991 [3]. Now MKS has been found in all age groups further strengthening the hypothesis that a congenital cause should be suspected. Some patients now are followed over a course of several years giving insights about the course of the disease and future prognosis for other patients. The symptoms reported haven't changed much since but diagnostics of the MKS and ability to help these patients have. The use of CT has enabled a better and easier visualization and determination of airway size, facilitating recognition of obviously abnormal cases. But as the airway measurements don't seem to be a routine assessment, it is not known how many cases just above the upper normal limit are missed. During the last 20 years, the new data about interventional strategies are the most important addition to the literature on MKS. Bronchoscopic treatment has developed quickly since 1991 and stenting now provides at least some relief to patients with MKS. Also surgical treatment has advance somewhat. And the fact that a clinical trial has been done, promises that evidence based recommendations may be available one day.

There is nothing much new that can be said about possible aetiology of MKS. Published cases still support the theory that MKS is a congenital defect but no new data has been published to better understand the cause of dilation. The authors of this review suspect it to be a multifactorial genetic effect, which causes the changes in airways with subsequent symptoms. The only publication on familial occurrence seems to be peculiarity and not a proof of heredity. Also the border between primary and secondary disease remains to be clarified.

The strength of this review lies in the comprehensive summary of previous publications about MKS, in the sheer number of cases reviewed and in the fact that the authors haven't limited their search only to English language. As the number of cases reported since the last review [3] has grown, so has the diversity of diseases encountered together with MKS, expanding experience of the treatment for these patients. But currently it is still impossible to say which cases are coincidental with MKS and if there are any conditions in which MKS is truly more common.

Some authors have assumed that in cases of rheumatologic and connective tissue diseases MKS should be considered to be secondary [4,8], [S96] but the authors of this review believe that it remains yet to be confirmed, as there are just few reports for each of the connective/rheumatologic diseases.

The authors of this review rebut a common misconception about the number of MKS cases published worldwide. The most common number mentioned is around 100 cases, which seems to be caused by careless citation; for example, an article [S97] stated this number in 2011, and cited a publication from 1994 [S42] which itself further referred to an article published in 1988 [S81]. A quick search in PubMed reveals more than 200 cases. The authors of this review estimate that there could be at least 300 cases published so far, if not more.

There are, of course, weaknesses to this review. First of all, it is based on case reports and lacks support of clinical trials. To the best knowledge of the authors every aspect of MKS has been included in this review but important details might have been missed, as articles that are not indexed in PubMed or Medline might still harbour important facts. Only what has been published so far can be reviewed and as seen from the example above — an important fact (successful lung transplantation) might not have been reported. The suspicion that this syndrome is under-reported only increases the doubt.

Data from trials are scarce and limited to specific patient groups. As the syndrome remains relatively rare and it is unlikely that any large scale systematic study will be conducted anytime soon, the authors of this review suggest that future publications should try to standardize reported clinical findings in order to decrease the wide variability observed in previous case reports. Patient's demographics (age, gender, and length of complaints, smoking status and pack years), clinical findings (including finger clubbing), spirometry values and bronchoscopic findings should be included. Both the transverse and sagittal measurements of trachea 2 cm above aortic arch, and the size of the left and right main bronchus should be given. This would eventually allow evaluating the characteristics of this syndrome more thoroughly.

For the treatment of MKS, the use of guidelines of non-cystic fibrosis bronchiectatic disease [29] and guidelines for lower respiratory tract infections [30] seem to be in place so that an appropriate antibiotic regiment upon exacerbations could be chosen. In the same time pathologic changes in airways caused by MKS may complicate the treatment of even community acquired pneumonia and may cause treatment resistance [23]. There is no data on prophylactic antibiotics use; therefore they should be applied according to the general guidelines. And finally, the authors suggest that vaccination for influenza and pneumococci should extend to all adult patients diagnosed with the condition regardless of their age or presence of symptoms.

Conflict of interest

Authors hereby acknowledge that they have no possible conflict of interest to report.

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Appendix A. Supplementary data

Supplementary bibliography designated with an "S" can be found on-line at http://dx.doi.org/10.1016/j.rmed.2013.08.042.

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