orphanaesthesia

Anaesthesia recommendations for

Mounier-Kuhn syndrome

Disease name: Mounier-Kuhn syndrome

ICD 10: Q32.4, Q32.1
with bronchiectasis: J47
exacerbation (acute): J47
lower respiratory infection: J47
acquired: J98.09
with bronchiectasis: J47
with exacerbation (acute): J47
with lower respiratory infection: J47.0

Synonyms: Tracheobronchomegaly, trachiectasis, tracheobronchomalacia, and multiple tracheal diverticula

Disease summary: Mounier-Kuhn syndrome (MKS) is an uncommon disease characterized by enlarged trachea and main bronchi, described anatomically as a particular condition of tracheobronchomegaly (TBM). It is also associated with repeated chest infections. This syndrome is considered to result by atrophy of the tracheal and bronchial elastic fibres, leading to thinning of the smooth muscles, tracheal flaccidity and dilatation and, subsequently, tracheobronchomalacia. Bronchiectasis are the most common comorbidity disorder associated with tracheobronchomalacia, but muscle layer weakness can also leads to the development of diverticula between the cartilaginous rings of the respiratory tree. The high rate of bronchiectasis combined with MKS strongly suggests that these pathological alterations are not restricted to the proximal airways but may involve the whole respiratory tract.

The main clinical manifestations of the syndrome are unspecific pulmonary symptoms, which widely vary from respiratory infections, chronic cough, and shortness of breath to a severe restriction of airflow in lung function tests.

The incidence of this syndrome is significantly higher in males. It is important to emphasize that tracheal enlargement does not progress over time; in fact, no correlation has been found between patients' age and airway diameter, and its severity evidently depends on other factors not yet determined.

An association of MKS has been found with some connective tissue diseases such as Marfan syndrome, Ehlers-Danlos syndrome, and Cutis Laxa. This suggests how the syndrome might have a genetic origin. However, to the current date no genetic mutation has been found.

Anaesthesiologist should suspect MKS whenever there is a lack of airway control due to the persistence of air leaks around a cuffed tracheal tube, when there is a discrepancy between the settled tidal volume and the volume detected during a mechanical lung ventilation or even if lung sounds are reduced on auscultation in both pulmonary fields.

Diagnosis requests the presence of the overall clinical, pathological, and radiological profile.

MSK is likely to be suspected when the diameter of the trachea and that of the major bronchi is greater than the 'normal' diameter as determined by a CT scan. Tracheomegaly is radiologically defined when tracheal diameter exceeds the upper limits of normality (25- or 21-mm coronal, 27 or 23 mm sagittal; in men and women, respectively). The upper limit of normal airway diameter is measured by examining the airway diameter of the normal healthy population without tracheobronchial disease and has conventionally been assumed to be mean + 3SD (standard deviation) in most studies.

Clinicians can perform an additional evaluation using fibre-optic bronchoscopy. Through bronchoscopy, it's possible to detect the dilatation of the trachea and proximal bronchi and in patients affected by tracheomalacia, expiratory airways collapse. Dynamic collapse of the trachea can be viewed by bronchoscopy and that is accepted as the gold standard in order to diagnose a condition of tracheomalacia. Additionally, the presence of diverticula on the posterior tracheal wall can be revealed.

Therapy is very minimal and only supportive and symptomatic. Pharmacologically, bronchodilators, inhaled corticosteroids and oral steroids may be helpful for many of these patients. Chest physiotherapy and pulmonary rehabilitation can enhance muco-ciliary clearance. Expiratory airway collapse and chronic hypercapnic respiratory failure can be managed through long-term oxygen therapy and non-invasive positive pressure ventilation. The use of positive pressure could preserve the patency of collapsed airways, acting as a 'pneumatic stent' to reduce the rate of airflow obstruction.

Moreover, this could possibly help the mobilization of bronchial secretions and improve lung ventilation.

Recurrent lung infections can be treated with antibiotics. Supportive measures include interventional rigid bronchoscopy, involving laser therapy or tracheal stenting.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Surgery is occasionally considered useful in the therapeutic management of Mounier-Kuhn syndrome due to the extensive nature of the disease. In end-stage disease, double lung transplantation has been reported, although it has given no demonstrated profit in terms of the likelihood of morbidity and mortality.

The anaesthetist's priority should be a correct airway management to ensure safety and adequate ventilation for the patient

Particularly, in case of lung transplantation, lung isolation is one of the greatest difficulties in the management of anaesthesia in a patient with MKS.

In these cases, the anaesthetist should consider various approaches such as extracorporeal membrane oxygenation or cardiopulmonary bypass to guarantee a sufficient tissue oxygenation and a metabolic status appropriate to the body's demands.

In a recent case report of sequential double lung transplantation, a backup cardiopulmonary bypass was used to achieve lung isolation and to aid the double lung transplantation.

A 10-mm single-lumen endotracheal tube was used to intubate the patient and the endotracheal cuff was filled with 25 ml of air to obtain minimal air leakage and an acceptable endotracheal cuff pressure (18-20 cm H_2O).

For the isolation of the right lung, a Fuji size 9 bronchial blocker was used, which was replaced many times, but total isolation was never realized. Thus, the blocker was removed, and the endotracheal tube was moved into the left main bronchus as an isolation strategy. The right lung was successfully grafted using this method.

The authors decided not to replace the endotracheal tube in the right main bronchus to avoid damaging the recently performed anastomosis of the same bronchus. Then, the surgeon blocked the left main bronchus with a vascular clamp to avoid an air leak and the left lung was grafted through a cardiopulmonary bypass.

Type of anaesthesia

According to the type of surgery.

Under general anaesthesia, pay close attention to the seal between the wall of the trachea and the inflated ET cuff. If the cuff is placed in an area of tracheal dilatation, the airway would not be secured, resulting in complications of ventilation.

In case of loco-regional anaesthesia in patients with Mounier-Kuhn syndrome, a recent case report recommends adequate preoperative diagnostic imaging of the vertebral spine to exclude dural ectasia. These assessments are recommended to be performed before undergoing lumbosacral spinal procedures to avoid an enhanced risk of failure of spinal anaesthesia or compromised epidural analgesia.

Necessary additional pre-operative testing (beside standard care)

CT scan, fibre-optic bronchoscopy.

From what can be deduced from the current data in the literature it is not necessary to perform tissue biopsies of the tracheal and bronchial wall to show a loss of elastic fibres.

Particular preparation for airway management

Tracheobronchomegaly should be assessed before surgery when general anaesthesia with endotracheal tube intubation is employed. Considerable peritubal leakage may be problematic, and inadequate ventilation may complicate the procedure.

A preliminary CT scan study is necessary to the measurement of the tracheal diameters, especially in the sections where the cuff of the endotracheal tube will be placed, to choose the size and the most suitable position for the tracheal tube.

Before induction, a set of different sized tubes, several supraglottic devices, a fiberscope, and a manometer to measure the pressure of the cuff should be prepared.

After endotracheal intubation, a small ET tube could be inserted in the oesophagus, cuffed and a stomach aspiration tube should be inserted into it to oesophageal tube, in order to reduce the risk of aspiration.

Particular preparation for transfusion or administration of blood products

According to type of surgery.

Particular preparation for anticoagulation

According to type of surgery.

Particular precautions for positioning, transportation and mobilisation

Not necessary and indicated.

Interactions of chronic disease and anaesthesia medications

Not reported.

Anaesthetic procedure

Before induction, a set of different sized tubes, several supraglottic devices, a fiberscope, and a manometer to measure the pressure of the cuff should be prepared.

A rapid sequence intubation should be performed in order to avoid gastric distension.

After endotracheal intubation, a small ET tube could be inserted in the oesophagus, cuffed and a stomach aspiration tube should be inserted into it to oesophageal tube, in order to reduce the risk of aspiration.

Care must be taken with ET cuff pressure because of the possible tracheal wall injury caused by further expansion of the tracheal tube cuff to avoid peri-tubal leakage.

A recent case in the literature reports airway management of a patient who was accidentally diagnosed with Mounier-Kuhn syndrome during general anaesthesia by sealing the oropharyngeal cavity with a wet gauze pack to obtain an adequate tidal volume even with a tube smaller than the tracheal diameter

If the ventilator detects an air loss, it is possible to place an extra-glottic device (I-gel, Fastrach) and a small ET tube through it, with the purpose of enhancing the seal thanks to the peri-laryngeal cuff.

Particular or additional monitoring

Pay particular attention to the pressure of the ET cuff to avoid mechanical stress on the tracheal wall.

Monitor strictly the Pressure/Volume loop on the ventilator in order to detect any air loss.

Possible complications

Air loss during ventilation.

Tracheal lesions or even rupture in case of excessive pressure from the cuff or during intubation.

Aspiration pneumonia.

Post-operative care

Strict chest X-ray monitor to detect early pulmonary complication from inhalation or atelectasis.

Early mobilisation and positional physiotherapy.

Prophylactic antibiotic therapy in case of respiratory infections.

Disease-related acute problems and effect on anaesthesia and recovery

Mounier-Kuhn syndrome should be considered in the differential diagnosis of spontaneous recurrent pneumothorax.

Ambulatory anaesthesia

See postoperative care.

Obstetrical anaesthesia

If possible, avoid general anaesthesia.

Tracheobronchomegaly acquired post COVID-19 infection

Acquired forms of “faux-MKS” have also been described in the literature.

Recent scientific publications suggest that severe COVID-19 is related to an increased incidence of tracheomalacia.

This complication is hypothesized to be a consequence of high-pressure ventilation for prolonged periods in those adult patients who require hyperinflation of endotracheal and tracheostomy tube cuffs to prevent peri-tubal cuff leaks. Hyperinflation of the cuff would lead to dilatation of the trachea and subsequent diagnosis of acute acquired tracheomalacia.

Tracheomalacia is commonly observed also in patients with a diagnosis of pulmonary fibrosis, in those undergoing prolonged mechanical ventilation, and in adults requiring prolonged hyperinflation of tracheostomy or endotracheal tube cuffs; all of these situations are commonly seen in patients with severe SARS-CoV-2.

Furthermore, all possible underlying co-morbidities, even rare ones such as MKS, should be carefully considered when managing a patient with COVID-19, because they may have a dramatic impact on the course of the disease.

These recent findings are particularly significant in the era of the COVID-19 pandemic because the eventual development of TBM as part of COVID-19 sequelae may lead to an increased risk of recurrent pulmonary infections.

In addition, this sort of tracheobronchomegaly may also have unexpected implications during general anaesthesia.

References

1. Cataldo R, Gallì B, Proscia P, Carassiti M. Management of a patient with Mounier-Kuhn syndrome undergoing repeated general anesthetics. Can J Anaesth 2013 Jun;60(6):602-3
2. I. Satia, B. Dua, N. Singh, K. Killian, and P. M. O'Byrne, "Tracheobronchomegaly, cough and recurrent chest infection: Mounier-Kuhn syndrome," ERJ Open Res., vol. 6, no. 2, pp. 00138–02020, 2020, doi: 10.1183/23120541.00138-2020
3. I. Satia, B. Dua, N. Singh, K. Killian, and P. M. O'Byrne, "Tracheobronchomegaly, cough and recurrent chest infection: Mounier-Kuhn syndrome," ERJ Open Res., vol. 6, no. 2, pp. 00138–02020, 2020, doi: 10.1183/23120541.00138-2020
4. E. Krustins, "Mounier-Kuhn syndrome: A systematic analysis of 128 cases published within last 25 years," Clin. Respir. J., vol. 10, no. 1, pp. 3–10, 2016, doi: 10.1111/crj.12192
5. https://www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=en&Expert=3347.
6. H.-J. No, J.-M. Lee, D. Won, P. Kang, and S. Choi, "Airway management of a patient incidentally diagnosed with Mounier-Kuhn syndrome during general anesthesia," J. Dent. Anesth. Pain Med., vol. 19, no. 5, p. 301, 2019, doi: 10.17245/jdapm.2019.19.5.301
7. A. Kuwal, N. Dutt, N. Chauhan, S. Kumar, S. Purohit, and L. K. Saini, "An Atypical Case of Mounier-Kuhn Syndrome: Time to Change the Diagnostic Approach?," J. Bronchol. Interv. Pulmonol., vol. 24, no. 1, pp. 84–87, 2017, doi: 10.1097/LBR.0000000000000226
8. S. Harper, M. Robinson, G. Manning, A. Jones, J. Hobson, and C. L. Shelton, "Management of tracheostomy-related tracheomegaly in a patient with COVID-19 pneumonitis," Anaesth. Reports, vol. 8, no. 2, pp. 159–162, 2020, doi: 10.1002/anr3.12076
9. C. C. Lee, B. S. Lin, J. Y. Chen, and C. C. Chuang, "Anesthesia for a patient with unexpected giant tracheobronchomegaly," Tzu Chi Med. J., vol. 29, no. 1, pp. 59–61, 2017, doi: 10.4103/tcmj.tcmj_1_17
10. A. G. Kaya, A. Çiledağ, Ç. Atasoy, and D. Karnak, "Flexible bronchoscopy and mechanical ventilation in managing Mounier-Kuhn syndrome: A case report," Sao Paulo Med. J., vol. 136, no. 3, pp. 266–269, 2018, doi: 10.1590/1516-3180.20160336270117
11. E. Lima et al., "Improving airways patency and ventilation through optimal positive pressure identified by noninvasive mechanical ventilation titration in mounier-kuhn syndrome: Protocol for an interventional, open-label, single-arm clinical trial," JMIR Res. Protoc., vol. 9, no. 8, pp. 1–7, 2020, doi: 10.2196/14786
12. B. Celik, S. Bilgin, and C. Yuksel, "Mounier-Kuhn syndrome: A rare cause of bronchial dilation," Texas Hear. Inst. J., vol. 38, no. 2, pp. 194–196, 2011
13. G. A. Sheridan, O. Godkin, and A. Devitt, "Dural ectasia associated with Mounier-Kuhn syndrome: A novel association in the context of spinal stenosis," BMJ Case Rep., vol. 2017, pp. 4–6, 2017, doi: 10.1136/bcr-2017-221439
14. N. E. Collins, "Acquired Tracheomegaly in Critically Ill Patients With COVID-19: A Literature Review," J. Nurse Pract., no. January, 2020
15. R. Jafari, L. Cegolon, F. Dehghanpoor, and M. Javanbakht, "Early manifestation of ARDS in COVID-19 infection in a 51- year-old man affected by Mounier-Kuhn syndrome," Hear. Lung, no. January, 202
16. S. Choudhury, A. Chohan, P. T. Taweesedt, R. Dadhwal, and A. Vakil, "Coronavirus Disease 2019-Induced Tracheomegaly: A Case Report," Cureus, vol. 14, no. 4, pp. 12–15, 2022, doi: 10.7759/cureus.23810
17. E. N. Unlu et al., "An unusual cause of recurrent spontaneous pneumothorax: The Mounier-Kuhn syndrome," Am. J. Emerg. Med., vol. 34, no. 1, pp. 122.e1-122.e2, 2016, doi: 10.1016/j.ajem.2015.05.05
18. Weissleder R, Wittenberg J, Harisinghani MG, et al. Primer of diagnostic imaging. Mosby Inc. 2007;ISBN:0323040683

19. Mcguinness G, Naidich DP, Leitman BS, et al. Bronchiectasis: CT evaluation. AJR Am J Roentgenol 1993;160(2):253-9
20. Cartier Y, Kavanagh PV, Johkoh T, et al. Bronchiectasis: accuracy of high-resolution CT in the differentiation of specific diseases. AJR Am J Roentgenol 1999;173(1):47-52
21. Barker AF. Bronchiectasis. N Engl J Med 2002;346(18):1383-93
22. Cantin L, Bankier AA, Eisenberg RL. Bronchiectasis. AJR Am J Roentgenol 2009;193(3):
23. W158-71
24. Solanki T, Neville E. Bronchiectasis and rheumatoid disease: Is there an association?
25. Br J Rheumatol 1992;31(10):691-3
26. Fenlon HM, Doran M, Sant SM, et al. High-resolution chest CT in systemic lupus erythematosus. AJR Am J Roentgenol 1996;166(2):301-7
27. Collins J, Stern EJ. Chest radiology, the essentials. Lippincott Williams & Wilkins 2007; ISBN:0781763142
28. Naidich DP, Srichai MB, Krinsky GA. Computed tomography and magnetic resonance of the thorax. Lippincott Williams & Wilkins 2007; ISBN:0781757657
29. Warrell DA. Oxford textbook of medicine, Sections 18-33. Oxford University Press, USA. 2005; ISBN:0198569785
30. Javidan-Nejad C, Bhalla S. Bronchiectasis. Thorac Surg Clin 2010;20(1):85-102
31. Javidan-Nejad C, Bhalla S. Bronchiectasis. Radiol Clin North Am 2009;47(2):289-306
32. Ooi GC, Khong PL, Chan-Yeung M, et al. High-resolution CT quantification of bronchiectasis: Clinical and functional correlation. Radiology 2002;225 (3):663-72
33. Mounier-Kuhn P. Dilatation de la trachee: Constatations, radiographiques et bronchoscopies. Lyon Med 1932;150:106–9
34. Spencer H. Congenital abnormalities of the lung; congenital tracheobronchomegaly. In: Spencer H, ed. Pathology of the Lung, 4th edition. Pergamon Press Oxford, 129–30
35. Aaby GV, Blake HA. Tracheobronchomegaly. Ann Thorac Surg 1966;2:64–70
36. Himalstein MR, Gallagher JC. Tracheobronchomegaly. Ann Otol Rhinol Laryngol 1973;82:223–7
37. Wolfgang Dahnert ED. Radiology Review Manual/second Edition, William and Wilkins 1993. Lippincot Williams and Wilkins NY, USA
38. Bateson EM, Woo-Ming M. Tracheobronchomegaly. ClinRadiol 1973;24:354–8
39. Sorenson SM, Moradzadeh E, Bakhda R. Repeated infections in a 68-year-old man. Chest 2002; 121:644–6
40. Giannoni S, Benassai C, Allori O, et al. Tracheomalacia associated with Mounier-Kuhn syndrome in the intensive care unit: treatment with Freitag stent. A case report. Minerva Anestesiol 2004;70: 651–9
41. Van Schoor J, Joos G, Pauwels R. Tracheobronchomegaly - the Mounier-Kuhn syndrome: report of two cases and review of the literature. Eur Respir J 1991;4:1303–6
42. Pilavaki M, Anastasiadou K, Vlachojanni E, et al. Tracheobronchomegaly (Mounier-Kuhn syndrome): Roentgen findings and tracheal stent instrumentation. Pneumologie 1995;49:556–8
43. Odell D, Shah A, Gangadharan S, et al. Airway Stenting and Tracheobronchoplasty Improve Respiratory Symptoms in Mounier-Kuhn Syndrome. Chest 2011;140:867–873.

Date last modified: **November 2022**

Authors:

Rita Cataldo, Anaesthesiologist, Department of Medicine and Surgery, Università Campus Bio-Medico di Roma, Roma, Italy, Research Unit of Anaesthesia, Intensive Care and Pain Management; Fondazione Policlinico Universitario Campus Bio-Medico, Operative Research Unit of Anaesthesia, Intensive Care and Pain Management¸ Roma, Italy
R.Cataldo@policlinicocampus.it

Alice Bellezze, Anaesthesiologist, Fondazione Policlinico Universitario Campus Bio-Medico, Operative Research Unit of Anesthesia, Intensive Care and Pain Management, Roma, Italy

Sabrina Migliorelli, Anaesthesiologist, Fondazione Policlinico Universitario Campus Bio-Medico, Operative Research Unit of Anesthesia, Intensive Care and Pain Management, Roma, Italy

Disclosure The authors have no financial or other competing interest to disclose. This recommendation was unfunded.

Reviewers:

Sudhakar Subramani, Department of Anaesthesia, University of Iowa, USA
sudhakar-subramani@uiowa.edu

Eduard Krustins, Department of Internal Medicine, Pauls Stradins Clinical University Hospital, Riga, Latvia
eduards.krustins@stradini.lv

Editorial review 2022

Tino Münster, Anaesthesiologist, Department of anaesthesiology and intensive care medicine, Hospital Barmherzige Brüder, Regensburg, Germany
Tino.Muenster@barmherzige-regensburg.de

Disclosure The reviewers have no financial or other competing interest to disclose.