orphananesthesia

Anaesthesia recommendations for patients suffering from

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

Synonyms: Tracheobronchomegaly, trachiectasis, tracheobronchomalacia, multiple tracheal diverticula

A marked dilation of the trachea and main bronchi, associated with a higher rate of recurrent lower respiratory tract infections, characterizes the Mounier-Kuhn syndrome (MKS). The clinical presentation varies widely from minimal respiratory discomfort, repetitive respiratory infections, mucus accumulation and ineffective cough, to severe respiratory failure. This syndrome is considered to be congenital, although most of the times it presents in the third or later decades with recurrent respiratory tract infections. The aetiology is uncertain, anatomo-pathological findings have led to believe it due to the lack of smooth muscle and elastic connective tissue in the trachea and main bronchi, causing tracheobronchomegaly and “herniation” of diverticula between the cartilaginous rings.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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Disease summary

Frequently the diagnosis can be sensed based on cases of abnormal chest X-ray, and an anaesthesiologist should be suspected it when there is an unexplainable loss of air during ventilation with tracheal tube, but it needs to be confirmed by a CT scan to allow for precise measurement of airways and evaluation of additional changes in pulmonary tissues, and possibly a bronchoscopy could be considered for additional evaluation.

The treatment does not include surgery and typically includes supportive measures and possible airway stenting in cases of significant tracheomalacia, but surgery has also been attempted. The only clinical trial which included 12 patients with Mounier-Kuhn syndrome showed significant improvements in pulmonary function and quality of life when compared to baseline. The objective of conservative treatment is sputum clearance, using positional physiotherapy and early and aggressive treatment of pulmonary infections. In some cases, chronic prophylactic administration of antibiotics may be required.

Typical surgery

Surgery is not generally indicated, but in some cases of severe tracheomalacia and expiratoy collapse of trachea a tracheobronchoplasty has been used, and in refractory end stage lung disorder lung transplant is the definitive treatment.

Type of anaesthesia

According to the type of surgery, lack of the “seal” in case of tracheal intubation should be concealed.

Necessary additional diagnostic procedures (preoperative)

CT scan, bronchoscopy.

Particular preparation for airway management

We suggest a thorough study of the CT scan and 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 the oesophageal tube, in order to reduce the risk of aspiration.

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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, transport or mobilisation

Not indicated.

Probable interaction between anaesthetic agents and patient's long-term medication

Not reported.

Anaesthesiologic 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 the oesophageal tube, in order to reduce the risk of aspiration.

Should the ventilator detect an air loss, it is possible to place an extra-gloctic device (I-gel, Fastrach) and a small ET tube through it, with the purpose of enhancing the seal with the oesophageal cuff.

Particular or additional monitoring

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

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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.

Postoperative 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.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease

None.

Ambulatory anaesthesia

See postoperative care.

Obstetrical anaesthesia

If possible, avoid general anaesthesia.

Literature and internet links

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