orphanaesthesia

Anaesthesia recommendations for patients suffering from

Inclusion body myositis

Disease name: Inclusion body myositis

ICD 10: M60.8

Synonyms: Sporadic inclusion body myositis

Inclusion body myositis (IBM) is a rare disease that is part of a group of muscle diseases known as idiopathic inflammatory myopathies, which are characterized by chronic excessive muscle inflammation associated with muscle weakness. IBM has a variable prevalence according to geographic, ethnic and age criteria. Prevalence in the general population ranges varies between 0.3 and 13.9 per 100,000.

IBM onset is usually over 50 years but may also occur earlier, in the 5th decade and it is more prevalent in males (2:1 over female).

First signs of the disease are usually weakness of the long finger flexors or quadriceps (typically flexor digitorum profundus and flexor pollicis longus). Alternative presenting phenotypes include dysphagia, foot drop or respiratory weakness. With the progression of the disease more groups of muscles are involved and the decrease of muscle strength is usually slowly progressive (occurs gradually over months or years). Dysphagia occurs in around 50% and can be severe, rarely requiring enteral feeding. Cardiac function appears to be spared in IBM. There are reports of sleep disordered breathing being identified primarily later in disease progression, although not necessarily correlated to severity of peripheral muscle weakness. The involvement of the respiratory muscles is characterized by a restrictive ventilatory syndrome of progressive evolution. Respiratory decline and dysphagia predisposing to pneumonia was the most common cause of death in a long-term follow-up of patients with IBM.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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

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Disease summary

The etiopathogenesis of IBM remains uncertain but muscle damage is considered to involve both an abnormal immune response reflected by excessive inflammatory infiltrates and a degenerative process resulting in rimmed vacuoles, mitochondrial impairment and protein aggregates. No causal gene has been identified but HLA-DR3 and 8-1 MHC genotypes have been shown to correlate with IBM susceptibility. Antibody cytosolic 5'-nucleotidase 1A, an autoantibody against the muscle protein cytosolic 5'-nucleotidase 1A, was detected in approximately half of patients but is also commonly seen in Sjogren's and SLE.

European Neuro Muscular Center published the ENMC IBM Research Diagnostic Criteria 2011, that include combined clinical and laboratory features: duration >12 months, age at onset >45 years, sCK no greater than 15 ULN and either knee extension weakness ≥ hip flexion weakness and/or finger flexion weakness >shoulder abduction weakness, or pathological features: endomysial inflammatory infiltrate, rimmed vacuoles, protein accumulation or 15-18nm filaments.

Differential diagnosis may include polymyositis and, in early stages of the disease, arthritis or motor neuron disease. There is no curative treatment for IBM, and patients usually do not respond to anti-inflammatory or immuno-modulatory therapies. Symptomatic treatments include exercise therapy and orthotic appliance. As the disease progresses, weakness can lead to frequent falls, walking aids and wheelchair use around 15 years after diagnosis. Bone fractures and other complications may occur as a result of falls. No change in mean life expectancy has been observed.

Typical surgery

Muscle biopsy, orthopaedic surgery (frequent bone fractures). Cases of severe dysphagia may require cricopharyngeal myotomy or placement of a gastrostomy tube.

Type of anaesthesia

Due to muscular weakness, if possible, regional anaesthesia is preferred but general anaesthesia need not be avoided if necessary. Regional anaesthesia avoids the need of artificial ventilation and the risk of associated respiratory complications in IBM patients. Some case reports with neuro-axial anaesthesia have been successfully described.

Necessary additional diagnostic procedures (preoperative)

Pulmonary function testing (including lung volumes and blood gas analysis) may be needed. An FVC is a useful screen for evidence of significant respiratory muscle involvement.

If regional anaesthesia is planned neurological consultation with a complete neurological exam is recommended for juridical reasons in certain countries.

Particular preparation for airway management

In advanced stages of the disease cricopharyngeal muscle weakness could increase the risk of pulmonary aspiration. Individual hospital prevention of pulmonary aspiration protocols should be considered.

Particular preparation for transfusion or administration of blood products

There is no evidence to support the need of particular anticoagulation. But the impaired mobility of advanced stage patients may suggest a higher risk of postoperative thrombosis.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transport or mobilisation

Not reported

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

Not reported. Provide steroid substitution in case of corticotherapy.

Anaesthesiologic procedure

The dose of nondepolarizing muscle relaxants should be reduced in order to avoid prolongation of muscle weakness. It is believed that succinylcholine should be avoided for the risk of hyperkalemia.

However, in a recent observational retrospective study with sixteen patients, Mortenson et al, objectified uneventful perioperative outcomes following general anesthesia with depolarizing and nondepolarizing muscle relaxants.

Particular or additional monitoring

If muscle relaxants drugs are used, monitoring muscular block is essential to avoid residual muscular block. With the recent introduction of Sugammadex® it's easier to ensure no residual block is absent.

Possible complications

Increased risk of aspiration pneumonia due weakness of the cricopharyngeal musculature.

Difficult or impossible safe extubation after general anaesthesia (due to residual effect of muscle relaxant or to the decrease in pulmonary function).

Postoperative care

If decreased in pulmonary function is found in postoperative care the use of inspiratory muscle training should be considered.

The assisted-cough techniques contribute to minimize the risk of respiratory complications to IBM patients throughout the perioperative period.

Reduced mobility may increase thromboembolic complication risk and should be minimised through active physiotherapy and provision of raised chairs and mobility aids to facilitate independent walking.

Ambulatory anaesthesia

Ambulatory surgery schedule may not be recommended because patient with more severe IBM should be monitored for a prolonged period after anaesthesia.

Obstetrical anaesthesia

The same recommendations of non-obstetrical anaesthesia should be used. Whenever possible, general anaesthesia should be avoided and regional anaesthesia used.

Literature and internet links

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This guideline has been prepared by:

Authors
Diana Vieira, Neusa Lages, Ana Marinho, Carlos Correia, Anaesthesiologists**,** Centro Hospitalar do Alto Ave, Hospital de Guimarães, Fafe, Portugal
lages.neusa@gmail.com

Peer revision 1
Stefen Brady, Neurologist**,** John Radcliffe Hospital, Oxford, United Kingdom
stefenbrady@nhs.net

Peer revision 2
James Miller, Neurologist, The Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, United Kingdom
james.miller@nuth.nhs.uk

Please note that this guideline has not been reviewed by two anaesthesiologsts but by two disease experts instead.