

orphan**a**nesthesia

Anaesthesia recommendations for patients suffering from **Welander distal myopathy**

Disease name: Welander distal myopathy

ICD 10: G71.0

Synonyms: late adult onset type 1 distal myopathy; distal myopathy Swedish type

Welander distal myopathy belongs to the group of distal myopathies. These are classified according to clinical features, inheritance pattern and histopathological criteria. Welander myopathy was first described by Lisa Welander in 1951. Inheritance occurs via autosomal dominant pathway. Welander myopathy has been linked genetically to the chromosome 2p13. This distal myopathy is almost exclusively found in Sweden and partly in Finland. Clinically, Welander myopathy has a late adult onset with slow progression with a mean age of onset of 45 years and a normal life expectancy. First symptoms appear as weakness combined with atrophy of distal muscles of the upper extremity leading to problems in small precision movements as well as the inability to extend the fingers. As the disease progresses the distal muscles of the lower extremity can be affected. Tendon reflexes are decreased or absent. Sensory dysfunction may occur in form of elevated thresholds for thermal stimuli in the distal parts of upper and lower extremity. Proximal involvement was described by Welander in very few cases and is therefore very rare and maybe just found in homozygous carriers. Cardiac involvement has been excluded. Creatine kinase values are either normal or slightly elevated. Nerve conduction velocities are normal. In electromyography both myopathic and neurogenic changes can be found. These include small polyphasic motor-unit potentials, reduced interference pattern, giant motor-unit potentials and spontaneous activity. Histopathological analysis shows increased variation of muscle fiber diameter, centrally located nuclei, split fibers, rimmed vacuoles as well as atrophic

Medicine in progress



Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Typical surgery

Muscle biopsy may be necessary in the course of the diagnostic process. There is no other typical surgery in patients with Welander distal myopathy.

Type of anaesthesia

General as well as regional anaesthesia can be applied equally.

There is no data indicating an advantage of either general or regional anaesthesia.

Necessary additional diagnostic procedures (preoperative)

In some cases, elevations of creatine kinase levels are reported. It is recommended to determine a preoperative baseline in case of perioperative complications such as rhabdomyolysis.

If regional anaesthesia is planned, a preoperative assessment of peripheral sensory nerve dysfunction should be considered.

Particular preparation for airway management

The disease affects only the distal and with some exception the proximal muscles of the upper and lower extremities. There are no reports concerning affection of oropharyngeal muscles as well as muscles involved in breathing. Thus, there should be no concerns with respect to airway management and respiratory complications.

Particular preparation for transfusion or administration of blood products

Not reported.

Particular preparation for anticoagulation

Patients with Welander distal myopathy underlie a slow progression of the disease without particular disability. Anticoagulation should be handled as in comparable patients with similar type of surgery without Welander distal myopathy.

Particular precautions for positioning, transport or mobilisation

Weakness of extensors in the lower extremity leads to some difficulties particularly in walking on the heels. Attention should be paid to patients in advanced states of Welander distal myopathy with pronounced weakness of distal muscles in the lower extremity.

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

Not reported. Patients with Welander distal myopathy do not take any particular chronic medication.

Anaesthesiologic procedure

There are no reports about patients with Welander distal myopathy undergoing either general or regional anaesthesia.

There are reports of patients with Welander distal myopathy suffering from restless legs syndrome. In these patients, application of propofol and etomidate should be avoided.

In all other patients, anaesthetics, opiates, and neuromuscular blocking agents can be used without concern.

As there are no reports about anaesthesia in Welander myopathy, there are no reports or concerns about malignant hyperthermia or the use of succinylcholine and volatile anaesthetics either.

Local anaesthetics can also be applied safely.

Particular or additional monitoring

Additional respiratory or cardiac monitoring is not needed.

However, neuromuscular monitoring should be judged carefully. Possibly, the neuromuscular response to stimulus may be diminished even before administration of neuromuscular blocking agents due to atrophy of peripheral muscles.

Possible complications

Welander distal myopathy does not affect the respiratory system. There are no reports on cardiac manifestation. One study analyzed autonomic cardiovascular responses in 9 patients with Welander distal myopathy. Normal respiratory sinus arrhythmia as well as heart rate response to vagal manoeuvres (Valsalva) was found. In orthostatic position, these patients had a greater increase of systolic pressure and reduced heart rate increase than in controls. The authors suggest that the peripheral vasomotor function might be altered in these patients.

In general, the probability for anaesthesiological complications is not higher than in a comparable healthy population.

Postoperative care

There is no data indicating that a particular postoperative care is needed.

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 diseases, e.g.:

Disease triggered emergency-like situations are not common in Welander distal myopathy.

Ambulatory anaesthesia

There is no particular data concerning ambulatory anaesthesia. We recommend adhering to common anaesthesiological guidelines.

Obstetrical anaesthesia

There is no specific data reported for obstetrical anaesthesia in female patients with Welander distal myopathy. Common guidelines should be respected.

Literature and internet links

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