orphanaesthesia

Anaesthesia recommendations for

Welander distal myopathy

Disease name: Welander distal myopathy

ICD 10: G71.0

Synonyms: Late adult onset type 1 distal myopathy

Disease summary: 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 as Myopathia distalis tarda hereditaria by Lisa Welander in 1951. Welander myopathy has been linked genetically to the chromosome 2p13 and inheritance occurs via autosomal dominant pathway. In a recent study Hackman et al. could identify a new mutation in TIA1 gene associated with Welander myopathy. This distal myopathy is almost exclusively found in Sweden and partly in Finnland. A German study found 3 patients with Welander phenotype in a total cohort of 42 patients with distal myopathy. Clinically, Welander myopathy has a late adult onset with slow progression with a mean age of onset of 45 years and 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 and ankle and brachioradialis reflexes are decreased or absent. Sensory dysfunction may occur in form of elevated thresholds for thermal and vibration stimuli in the distal parts of upper and lower extremity. Proximal muscle involvement was described by Welander in very few cases with advanced disease or in severe homozygous cases. Cardiac involvement has been excluded. CK 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 fibers.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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Typical surgery

Muscle biopsy can be necessary for diagnosis. There is no other typical surgery in patients with Welander distal myopathy.

Type of anaesthesia

There is no literature concerning anaesthesiological risks in patients with Welander distal myopathy. There are no reports for cardiac involvement in Welander myopathy. Therefore, evaluation for cardiovascular risk should be performed as in other healthy patients on the basis of international guidelines. However, there is no data in this concern. CK values should be determined before anaesthesia and surgery in order to have a baseline in case of perioperative rhabdomyolisis.

Necessary additional pre-operative testing (beside standard care)

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 rhabdomyolsis.

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, transportation and mobilisation

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

Interactions of chronic disease and anaesthesia medications

Patients with Welander distal myopathy do not take any particular chronic medication that may interact with anaesthetic agents.

Anaesthetic procedure

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

There are reports of patients with Welander distal myopathy suffering 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.

There is no correlation between malignant hyperthermia and Welander distal myopathy in literature. The use of succinylcholine and volatile anaesthetics can be considered as safe.

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 orthosthatic 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.

Post-operative care

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

Disease-related acute problems and effect on anaesthesia and recovery

Disease triggered emergency-like situations concerning anaethesia are not reported in Welander distal myopathy.

Ambulatory anaesthesia

Not reported.

Obstetrical anaesthesia

Not reported.

References

1. Ahlberg G, Jakobsson F, Fransson A, Moritz A, Borg K, Edström L. Distribution of muscle degeneration in welander distal myopathy – a magnetic resonance imaging and muscle biopsy study. Neurmusc Disord 1994;4(1):55–62
2. Ahlberg G, Borg K, Edström L, Anvret M. Welander distal myopathy is not linked to other defined distal myopathy gene loci. Neuromusc Disord 1997;7:256–260
3. Borg K, Sachs C, Kaijser L. Autonomic cardiovascular responses in distal myopathy (Welander). Acta Neurol Scand 1987;76(4):261–266
4. Borg K, Ahlberg G, Anvret M, Edström L. Welander distal myopathy – an overview. Neuromuscular Disorders 1998;8:115–118
5. Borg K, Ahlberg G, Borg J, Edström L. Welander's distal myopathy: clinical, neurophysiological and muscle biopsy observations in young and middle aged adults with early symptoms. Journal of Neurology, Neurosurgery and Psychiatry 1991;54:494–498
6. Borg K, Borg J, Lindblom U. Sensory involvement in distal myopathy (Welander). Journal of the Neurological Sciences 1987;80:323–332
7. Dimachkie M and Barohn R. Distal Myopathies. Neurol Clin. 2014; 32(3):817–842
8. Illa I. Distal myopathies. J Neurol 2000;247:169–174
9. Hackman P, et al. Welander distal myopathy is caused by a mutation in the RNA-binding protein TIA1. 2013 Ann Neurol 73(4):500–509
10. Kraya T and Zierz S. Distal myopathies: from clinical classification to molecular understanding. J Neural Transm 2013;120(Suppl 1):S3–S7
11. Nonaka I. Distal myopathies. Current Opinion in Neurology 1999;12(5):493–499
12. Von Tell D, Somer H, Udd B, Edström L, Borg K, Ahlberg G. Welander distal myopathy outside the Swedish population: phenotype and genotype. Neuromusc Disord 2002;12:544–547
13. Welander L. Myopathia distalis tarda hereditaria. Acta Med Scan 1951;141(Suppl 265):1–124.

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This recommendation was prepared by:

Author(s)

Doris Rohde, Anaesthesiologist, University Hospital Erlangen, Germany
doris.rohde@kfa.imed.uni-erlangen.de

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This recommendation was reviewed by:

Reviewer 1

Tino Münster, Anaesthesiologist, University Hospital Erlangen, Germany

Reviewer 2

Lars Edström, Neurologist, Karolinska-Institute, Stockholm, Sweden
lars.edstrom@ki.se

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

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