Palliative care in neuromuscular diseases.

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Abstract

PURPOSE OF REVIEW: Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness. Neuromuscular disorders (NMDs) are characterized by progressive muscle weakness, leading to pronounced and incapacitating physical disabilities. Most NMDs are not amenable to curative treatment and would thus qualify for palliative care. Amyotrophic lateral sclerosis is a relentlessly progressive disease, which leads to death about 2 years after onset due to respiratory muscle weakness. Increasingly, neurologists caring for these patients learn to apply the principles of palliative care. However, this does not yet apply to other well known and frequently occurring NMDs.

RECENT FINDINGS: There is sparse literature on palliative care in NMDs such as Duchenne muscular dystrophy, spinal muscular atrophy, muscular dystrophies, some congenital myopathies, Pompe's disease and myotonic dystrophy type 1. These NMDs are often associated with imminent respiratory insufficiency and/or heart failure leading to a reduced life expectancy. Reasons for underutilization may include misconceptions about palliative care amongst patients, family carers and healthcare professionals or lack of awareness of the usefulness of this approach in these severely affected patients and the possibilities of integration of palliative principles into care for children and adults with NMDs.

SUMMARY: There is an urgent need for increased attention to the development of palliative care in chronic progressive neuromuscular diseases associated with increasing functional incapacities and premature death. This will require education and training of the healthcare professionals, involvement of patient associations and funding to perform research.

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