Report on 228th ENMC International Workshop:

AIRWAY CLEARANCE TECHNIQUES IN NEUROMUSCULAR DISORDERS

3 – 5 March 2017

Acknowledgements:

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On 3 March 2017, 21 participants from 12 countries across the world (including Australia, Europe; United Kingdom; United States of America; and South Africa) met in Naarden, Netherlands, to attend and contribute to the 228th ENMC International Workshop entitled, "**Airway Clearance Techniques In Neuromuscular Disorders".** Participants represented clinicians, researchers, patients and patient advocacy groups, from a range of socioeconomic and geographical backgrounds and disciplines. The workshop was conducted under the leadership of Michel Toussaint (Belgium); David Berlowitz (Australia); Jesus Gonzalez (France) and Michelle Chatwin (United Kingdom). Professional participants for this ENMC Workshop were selected for their active involvement in neuromuscular disorder (NMD) research, clinical care, advocacy, policy development and/or education, and are all considered experts in their respective areas.

Workshop rationale and scope

There are a number of congenital and acquired NMDs, which may affect the respiratory muscles, to varying degrees and at different life stages (from infancy, through childhood, and to adulthood). This weakness results in patients not being able to effectively cough and clear pulmonary (lung) secretions on their own, leading to recurrent chest infections and ultimately, chronic changes in the lungs. Airway clearance techniques (ACTs), to facilitate clearance of

pulmonary secretions, along with ventilatory support as needed, are therefore essential components in the care of people with NMDs and associated respiratory muscle weakness.

Currently, national and centre- specific practices vary substantially and adherence to published guidelines for the management of secretion encumbrance is not optimal. This workshop was therefore conducted with the primary aim of developing a consensus guideline for the minimum and optimal standards of ACT care for patients with a range of NMDs and clinical presentations, from childhood through to adulthood.

The following major topics were discussed during the workshop proceedings:

1. The pathophysiology of secretion encumbrance in people with NMDs.
2. The principles of proximal and peripheral airway clearance.
3. The effect of respiratory tract infection on respiratory muscles, lung volumes and blood gas exchange in people with NMDs.
4. Detailed description of all peripheral and proximal ACTs available for use in patients with weak respiratory muscles.
5. The limits of effectiveness of each ACT, the need for a learning period/training, financial cost, availability and possible complications.
6. Specific treatment algorithms (protocols) were defined for ACT management in NMDs in terms of: context, patient age and ability to cooperate, presence of invasive or noninvasive device interface, glottic control and objective measures of respiratory muscle strength.
7. Optimal outcome measures for future research studies were identified.

The workshop generated enthusiastic discussion from all participants and achieved its stated objectives, with a high level of agreement for all final recommendations reached.

The full report (to be published in the journal *Neuromuscular Disorders*) will provide practical recommendations that can be used by physicians, respiratory therapists/physiotherapists, patients and their carers. In addition, a state of the art review of ACT in patients with NMDs will be published. These publications will provide a novel framework for practical implementation in different clinical settings, as well as identifying priority areas for research. By optimising ACT management, it is hoped that duration and quality of life will be optimised, and the number of pulmonary exacerbations requiring hospitalisation and invasive ventilation be reduced in people with NMDs throughout the world.