Shedding light on oropharyngeal dysphagia in myotonic dystrophy type 1

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Valorization
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Valorization is defined as “the process of creating value from knowledge, by making knowledge suitable and/or available for social (and/or economic) use and by making knowledge suitable for translation into competitive products, services, processes, and new commercial activities” (National Valorization Committee). The studies presented in this thesis were intended to help determine the characteristics of oropharyngeal dysphagia (OD) in myotonic dystrophy type 1 (DM1), and to understand and subsequently improve the clinimetric characteristics of the fiberoptic endoscopic evaluation of swallowing (FEES) scales.

OD, which involves difficulty with eating and drinking, is a disorder that affects over 16 million people in the USA and over 40 million in Europe. It is a common symptom in neurological diseases such as stroke, Parkinson’s, Alzheimer’s, and DM1. Its complications may include malnutrition, dehydration, and aspiration pneumonia; in DM1 patients, pneumonia is reported as one of the most frequent primary causes of mortality.

OD also takes a heavy financial toll on the healthcare system. In 2010 the economic impact of dysphagia in hospital settings in the USA was calculated to be USD 547 million annually. That financial burden is linked to the extended length of hospitalization, higher rate of respiratory infection, increased expenditure on medical equipment and treatment, e.g. enteral feeding, antibiotics, etc., that are associated with dysphagia. While the cost of treating OD can be quantified, the social and psychological burden that significantly reduces quality of life for both patients and caregivers is more difficult to evaluate. Until 2010, there were 1117 patients with myotonic dystrophy registered in the Netherlands. The exact prevalence of OD in these population is not known; the literature worldwide puts it in the range of 25 to 80%. Despite the importance of preventing its associated complications, OD is often under-diagnosed, presumably because it is overlooked in the clinical assessment of DM1 patients.

Professionals from several disciplines are involved in the assessment and management of dysphagia: medical specialists, speech-language pathologists, nurses, dieticians, and caregivers. Because dysphagia requires a multidisciplinary approach, the results of our research are of relevance to all these professions. Therefore, we recommend a multidimensional assessment to evaluate swallowing in DM1. The underlying idea is that early and precise identification of OD may prevent complications that would reduce personal health-related quality of life and increase healthcare costs.

Diagnostics and treatment of OD in DM1 frequently rely on low to moderate levels of evidence, as compared to the literature available to cardiologists or neurologists, for instance. In our opinion, the paucity of well-designed studies in dysphagic DM1 patients cannot be attributed to disinterest on the part of patients as well as the medical community. Rather, the current state of knowledge apparently reflects the scarcity of financial resources for basic research in this field, as most of governmental and private-sector funding favors research with societal and economic applications. We do not
dispute the importance of knowledge valorization, i.e., making knowledge suitable or available for social and/or economic use. Yet we do question the feasibility of evidence-based practice in the healthcare and medical sectors without support from basic and fundamental research.

The studies presented in this thesis do not directly address the implementation of an assessment procedure or a specific treatment modality for OD. All our studies could be classified as ‘basic or exploratory research’ and, as such, their societal impact is more difficult to establish. We do believe, however, that in the longer term they will serve as stepping stones, paving the way for future studies on the implementation of diagnosis and/or disease-specific guidelines for identification of OD in DM1 patients. A more refined understanding of the psychometric characteristic of FEES scales, of the underlying pathophysiology of dysphagia in DM1 as well as of patients’ perception of the swallowing impairment are fundamental to the development of disease-specific guidelines and different treatment modalities specifically for this population.
References
