High nutritional risk in patients with oculopharyngeal muscular dystrophy: association with dysphagia

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Oculopharyngeal muscular dystrophy (OPMD) is a rare hereditary neuromuscular disease with the highest worldwide prevalence in Québec (Canada). As OPMD progresses over time, patients present with an atrophy of the pharyngeal muscles and therefore, swallowing problems (dysphagia) that appear around 40-60 years old. Dysphagia can compromise food intake and ultimately, patients’ nutritional status. Moreover, these patients present limb weakness that can lead to functional limitations, such as grocery shopping, which can contributes to the risk of poor nutrition. To date, no study has investigated the nutritional risk of OPMD patients. Objective: To assess the nutritional risk in adults with OPMD in association with oropharyngeal dysphagia. Methods: In this cross-sectional study, 40 adults aged 50-75 years and with molecular confirmation of OPMD are currently recruited among patients of a neuromuscular rehabilitation clinic. Nutritional risk is assessed with the French version of the Seniors in the Community: Risk Evaluation for Eating and Nutrition II (SCREEN II) and the severity of dysphagia with the French-Québec version of the Sydney-Swallow Questionnaire (SSQ). Anthropometric data are also measured using standardized procedures. Preliminary results: Data are now available for 20 patients aged 51 to 73 years old; 10 (50%) being men. Mean BMI (±SD) is 25.8±4.1, with 3 (15%) patients having low BMI and only one (5%) reporting weight loss >2.5 kg over the last 6 months. SCREEN II scores showed high nutritional risk in 17 (85%) of OPMD patients. SCREEN II items contributing the most to the nutritional risk scores are swallowing difficulty (75%), consumes daily 3 fruits and vegetables (70%) and 1-2 milk products (70%) or less, limits foods (55%), skips meals (50%) and chewing difficulty (50%). Pearson’s correlation coefficient showed significant association between dysphagia severity and nutritional risk (r = -.456 p =.043). Conclusion: To our knowledge, this ongoing study is the first to investigate the nutritional risk in OPMD. Our preliminary data indicate that individuals with OPMD present high nutritional risk, without presenting low BMI. The present study highlight the need for dietary counseling in these patients. [Funded by the Association française contre les myopathies and the Fondation de ma vie.]