Methods. – Sixteen FSHD patients randomized (trained/controls) were recruited in the Rhône-Alpes Reference Centre for Rare Neuromuscular Diseases consultations. The training group realized three weekly sessions on cycle. Multi-factorial evaluations, before, after and over the training period were performed in both groups: maximal strength, fatigue resistance, aerobic power, questionnaires of quality of life and functional tests. Biopsies from vastus lateralis muscles were done (beginning and end of the program). Program started under a coach supervision and was monitored by heart rate recordings.

Results. – Patients performed this program of exercise therapy with a great assiduity and an excellent tolerance. We report multi-factorial benefits of this home-based training program and its influence on the quality of life of trained patients.

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Rasch analysis of the motor function measure in patients with congenital muscle dystrophy and congenital myopathy

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Keywords: Dystrophie musculaire congénitale; Myopathie congénitale; Évaluation de la fonction motrice; Mesure des résultats; Analyse Rasch

Objectives. – Valid outcome measures are necessary to monitor the treatment effects in patients with congenital disorders of muscle.

Methods. – In 19 departments in France, Belgium, and USA, 289 patients aged 5- to 77-years-old were enrolled. A Rasch analysis examined the robustness of the motor function measure across the disease spectrum. The three domains (standing position and transfers, axial and proximal motor function, and distal motor function) were examined using RUMM 2030 software with a partial credit model.

Results. – The original 32-item MFM did not fit the Rasch model expectations enough in neither of its domains. Switching from a four- to a three-category response-scale in 18 items restored response order in 16. Various additional checks suggested the removal of seven items. The resulting 25-item MFM demonstrated a good fit to the Rasch model. Domain 1 was well-targeted to the whole severity spectrum whereas domains 2 and 3 were better targeted to severe cases. The reliability coefficients MFM-25 suggested sufficient ability for each summed score to distinguish between patient groups (0.9, 0.9, and 0.7 for domains 1, 2, and 3, respectively).

Discussion. – The Rasch-scaled MFM-25 can be assumed to be a linear scale in each of its three domains.

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