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## Use of Fatigue Index as a Measure of Local Muscle Fatigability in Ryanodine Receptor Isoform-1-Related Myopathies.

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### Abstract

**Introduction:** Individuals affected with ryanodine receptor isoform-1-related myopathies (*RYR1*-RM) commonly experience fatigability in the quadriceps, which may limit physical function and potentially diminish quality of life. Fatigability, in *RYR1*-RM, results from skeletal muscle injury secondary to dysfunction of the major skeletal muscle  $Ca^{++}$  channel. However, during fatigability testing, affected individuals did not always reach the point of local muscle fatigue as defined by a fatigue index (FATI) at 50% of peak torque. Surakka et al. compared three versions of FATI equations, which vary by the area under the force curve (AUC). By performing this comparison, they were able to determine the optimal equation in individuals with Multiple Sclerosis. **Purpose:** Using a similar comparison, we sought to identify the optimal FATI equation in the *RYR1*-RM population. Secondly, because local muscle fatigability might have an impact on independent living, this study also assessed change in local muscle fatigability over a 6-month time frame. **Methods:** Thirty participants were analyzed from the *RYR1*-RM natural history study and double-blind, placebo-controlled N-acetylcysteine (NAC) trial, [NCT02362425](#). Twenty-seven had fatigability data, from isometric knee extension and flexion fatigability tests, available for the purpose of establishing a method for predicting FATI at 50% peak torque. For the natural history study, 30 participants were used to assess disease progression of local muscle fatigability achieved during the knee extension fatigability test, and 29 participants for the knee flexion fatigability test. **Results:** Surakka's equation 1, using the prediction approach, led to the smallest median error, the smallest square-root of uncorrected sum of squares, and the smallest average of the absolute value of the differences. No difference was observed in FATI at 50% peak torque between month 0 and month 6 for extension ( $p = 0.606$ ) and flexion ( $p = 0.740$ ). **Conclusion:** Surakka's equation 1, with the prediction approach, was found to be the most accurate for imputing values when fatigue was not reached during a sustained knee isometric fatigability test in *RYR1*-RM.

Furthermore, when used to assess fatigability-based disease stability, local muscle fatigability, in this RYR1-RM population remained stable.

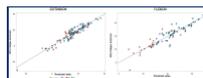
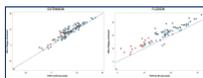
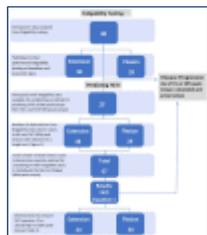
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**KEYWORDS:** RYR1-RM; fatigability; fatigue index; muscle; neuromuscular

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