The COVID-19 pandemic disrupted countless human activities since 2020. In most places, face-to-face visits for research projects were interrupted for months; some studies were adapted to use remote methods. One challenge was to find an instrument prone to remote application that could detect a reduction in functional status among carriers of spinocerebellar ataxias. The most used criterion is the cutoff of 3 points in the Scale for Assessment and Rating of Ataxia (SARA) [1]. The remote SARAhome version was recently proposed [2], but the technology needed was not available in research centers like ours yet.

As an alternative, we studied the Friedreich Ataxia Rating Scale/activities of daily living (FARS-ADL) [3], a patient-reported outcome designed to evaluate limitations in functional status of ataxic subjects.

We aimed to test if FARS-ADL could distinguish subjects with SARA score  \( \geq 3 \) (ataxics) among persons belonging to families with spinocerebellar ataxia type 3/Machado-Joseph disease (SCA3/MJD). Confirmed carriers and their relatives at 50\% risk were evaluated during July and August 2021. FARS-ADL was administered as a structured interview by telephone; SARA and DNA samples were collected simultaneously in our institution within 15 days after FARS-ADL. Participants at 50\% risk and examiners were kept blinded to their genetic results.

Nineteen ataxic (with SARA  \( \geq 3 \)) and 13 pre-ataxic (with SARA  \( < 3 \)) SCA3/MJD carriers, and 13 related controls were included, with median (IQR) ages of 44.01 (19.00), 29.00 (8.00), and 40.00 (13.75) years. Ataxic and pre-ataxic subjects carried 75 (4) and 75 (4) CAG repeats in their expanded alleles; age at onset of gait ataxia was 43 (19) in ataxics. Two alternatives were used to define FARS-ADL cutoffs between ataxic and pre-ataxic. According to a maximum-accuracy cut-point, FARS-ADL values less than 4 detected persons with SARA  \( < 3 \), while values greater than 8 detected persons with SARA  \( \geq 3 \) (Fig. 1A). According to the ROC curve and Youden’s index, a FARS-ADL score larger than 4 points detected presence of SARA  \( \geq 3 \), with 7.7\% and 5.6\% of false-positives and false-negatives, and with sensitivity and specificity of 0.94 and 0.92.

Former reports on simultaneous FARS-ADL and SARA data were obtained in Friedreich’s Ataxia (FRDA) subjects with SARA larger than 3 [4, 5], where 57 out of 594 subjects...
showed FARS-ADL scores of 4 or less (Reetz, personal communication). This would be equivalent to 9.6% false-negatives if FARS-ADL larger than 4 was used to classify FRDA subjects as ataxic.

Remote evaluations of persons at risk for ataxia might continue to be a demand for the near future. FARS-ADL is an easy to perform questionnaire through online interfaces or telephone calls. FARS-ADL does not detect ataxia, but in this population at risk for ataxia, it might help investigators to assume which subjects are already ataxic in the temporary impossibility of using the gold-standard method SARA. The specificity was high in SCA3/MJD, but not sufficient for FRDA. Therefore, it will be important to study more pre-ataxic and ataxic carriers to confirm the usefulness of FARS-ADL as a remote predictor of the symptomatic/ataxic status in SCA3/MJD and in other forms of ataxia.

Acknowledgements We thank all the subjects and families who contributed to this study.

Authors’ Contributions ECM contributed to the conception, organization, and execution of research project; helped in review and critique statistical analysis and critically revised the manuscript; and helped in manuscript review and critique. VHIS, CMO, and GB contributed to the research project execution, statistical analysis review and critique, and manuscript review and critique. MLSP contributed to the research project execution and manuscript review and critique. VLT contributed to the research project conception and execution, statistical analysis design and execution, and manuscript review and critique. LBJ contributed to the research project conception and organization; helped in the design, and review and critique of the statistical analysis; helped in the writing, review, and critique of the first draft of the manuscript. All authors read and approved the final manuscript.

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