Cortico-kinematic coherence is less sensitive to disease progression than an ataxia score in Friedreich ataxia

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General Submission

My Abstract

Friedreich ataxia (FRDA) is a rare genetic disorder caused by pathologic expansion of an intronic GAA repeat which disrupts transcription of frataxin, a mitochondrial protein. It is mainly characterized by slowly progressive cerebellar and afferent ataxia. Neurological deterioration can be captured by clinical rating scales such as the Scale for Assessment and Rating of Ataxia (SARA). Most SARA items are assessed under visual control, making the scale more sensitive to cerebellar than to afferent proprioceptive components of ataxia. Cortico-kinematic coherence (CKC), which is mainly driven by movement-related afferent proprioceptive inputs to contralateral primary sensorimotor (SM1) cortex, is decreased in both passive and active movements in FRDA patients. CKC, which indexes the coupling between cortical activity and hand movement kinematics during repetitive finger movements, typically peaks at movement frequency (F0) and its first harmonic (F1) at contralateral SM1 cortex. This prospective and longitudinal magnetoencephalography (MEG) study assesses if CKC is able to track the neurological deterioration observed in FRDA patients over a one-year period.

CKC was evaluated using whole-scalp-covering MEG (Neuromag Vectorview & Triux, Elekta Oy) in 12 right-handed FRDA patients (8 females, mean age: 26 years, range: 9-46 y; mean SARA score: 20.3, range 4.5-25.5) in two sessions performed at about one-year interval (mean 12.75 months, range 10-15 months). CKC was evaluated both at movement frequency (F0) and its first harmonic (F1) during active right forefinger-thumb opposition movements (Active, aCKC) and during passive right forefinger flexionextensions generated by an artificial muscle stimulator (Passive, pCKC). Forefinger acceleration was monitored with an accelerometer. Movement regularity was estimated for aCKC as the full width at half maximum of the accelerometers spectral peak at F0 (e.g., the wider, the less regular). Sensor-space CKC was calculated for

rolandic sensors contralateral to the movements. SARA score, CKC and movement regularity values between the two sessions were compared with two-tailed paired t-tests.
SARA score significantly deteriorated between session 1 and 2 (20.3 +/- 7.7 SD vs 22.04 +/- 6.1 SD, p=0.035) as did movement regularity (18%; p=0.014). However, no significant difference was found between CKC values at F0 or F1 between the two sessions, both for Active (F0, 0.13 ± 0.07 vs 0.11 ± 0.12, p=0.6; F1: 0.09 ± 0.13 vs 0.08 ± 0.07, p=0.69) and Passive (F0, 0.1 ± 0.06 vs 0.1± 0.1, p=0.9; F1, 0.06 ± 0.08 vs 0.04 ± 0.07, p=0.6) conditions.
This study shows that CKC is less sensitive to FRDA-related neurological deterioration than the SARA score. This finding might be related to the fact that FRDA progression is due to a combination of progressive proprioceptive, cerebellar and corticospinal dysfunction, while CKC mainly is mainly driven by afferent proprioceptive inputs to contralateral SM1 cortex. This finding suggests a lack of CKC sensitivity to detect subtle FRDA-related proprioceptive tracks degeneration.