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Detection of an Ataxia-type disease from EMG and IMU sensors

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**Abstract**—Mitochondrial recessive ataxia syndrome (MIRAS) is a heritable disease, relatively common in Finland. Among other things, patients suffer from ataxia, a movement disorder with difficulties in coordination. To date, no treatment is known for the disease, but medication and therapy can lessen the symptoms, provided that the progression of symptoms is closely monitored to adjust the treatment according to the individual needs. This necessary evaluation is a manual, subjective process.

We report about our efforts to explore quantifiable characteristics that could be used to monitor the disease progression objectively using electromyography (EMG) as well as inertial measurement unit (IMU) sensors. In particular, in a study with eight participants, including a patient, we have collected muscle activation as well as IMU data during several tasks. The study found some characteristics that might qualify as indicators of ataxia, such as high-frequency electrical activity (EA) components and similarity of repetitions. We further suggest the use of IMU and machine learning to improve the objective monitoring of the disease's progression.

**Index Terms**—MIRAS, Ataxia, sEMG, IMU

I. INTRODUCTION

Mitochondrial recessive ataxia syndrome (MIRAS) is the most common genetic ataxia disease in Finland, caused by a mutation in the polymerase gamma-gene (POLG1) and with a high carrier frequency in the general population of 1:125 [1]. The disease was first discovered in adult patients at the beginning of the 21st century as an adulthood ataxia-neuropathy. After the initial discovery, the clinical phenotype and onsetting age have broadened with further studies. The onsetting age varies from 0 to 50-year-olds, and the phenotypes can be categorized based on the onsetting age since there are distinguished symptoms. The three identified clinical phenotypes are childhood-, juvenile- and adult-onset MIRAS. The childhood onset is often referred to as Alpers or Alper-Huttenlocher syndrome. The juvenile-onset MIRAS includes common symptoms of refractory epilepsy and migraine-like headaches. The most common form of MIRAS phenotypes in Finland is the adult-onset phenotype, which comprises two-thirds of MIRAS patients. The common symptoms for the adult-onset MIRAS are ataxia, neuropathy, psychiatric symptoms, and cognitive impairment [1], [2]. Many of the mechanisms behind this disease are still unknown [2]. Currently, MIRAS can only be treated for symptoms, i.e., with palliative care, meaning that the progression of the disease cannot be influenced with therapy.

Since little is known about the disease, it is common that patients try new medications and doses, which in turn demands frequent and lengthy visits to a medical expert, which is burdening for the patient and the expert. Furthermore, due to the constrained time during the visits, the analysis might be less comprehensive than desired and is further affected by the specific form of a day. Likewise, the reports of the patients can be very subjective and thus hinder optimal treatment.

Technical support, which would help a patient to track the impact of medications on the progress of the disease, may relieve part of this burden when hospital visits can be reduced in exchange for remotely monitored tests. In the frame of this work, we investigated the use of electromyography (EMG) and inertial measurement units (IMU) during different arm movements to analyze, monitor, and describe objectively conditions of the patients that may indicate MIRAS and its progression. In particular, EMG allows to measure the activity of muscles during different tasks and could therefore help identify underlying issues causing movement difficulties for the patients. We hope that, prospectively, a patient may be empowered through these findings by conducting a set of (guided and normalized) exercises to collect data on the condition and on the progress of the disease. This article develops different tasks, uses surface EMG sensors to record muscle activities, and seeks approaches to distinguish MIRAS patients from people not affected by the disease by analyzing EMG and IMU data through different tasks.

II. BACKGROUND AND RELATED WORK

MIRAS was first described by a research team at the University of Helsinki in 2005 [1]. To date, the state of the disease and necessary treatments are evaluated manually through regular meetings with a medical expert. In particular, the physician interprets the patient’s self-assessment and evaluates the patient’s performance on defined tasks. To assess the progress of the disease, different standardized tests, as well as rating procedures and scales, are used. Examples are the International Cooperative Ataxia Rating Scale (ICARS) [3], the Scale for the assessment and rating of ataxia (SARA) [4], or the Friedreich Ataxia Rating Scale (FARS) [5], [6].

1https://neuroriitto.fi/tieto-tuki/tietoasairuksista/harvinaiset-neurologiset-sairaudet/diagonoosit/miras/
Ataxia is a degenerative disease of the nervous system, consisting of a lack of voluntary coordination of muscle movements. Therefore, it might be possible to diagnose the state of the disease by analyzing muscle activity, for instance, using electromyography (EMG), the measurement of the electrical activity (EA) of a muscle. In particular, in a common, bipolar approach, two electrodes are placed on the skin near the muscle belly along the muscle fibers. The EA of the muscle fibers under the skin is recorded as a voltage difference between the electrodes. Specifically, surface electromyography (sEMG) refers to the measurements in which a muscle’s EA is recorded by electrodes attached to the skin above the muscle. Since the raw signal still contains random components, the root-mean-square (RMS) of short windows of the signal is commonly calculated as a measure for the EA. The median frequency (MF) of the raw sEMG signal is another parameter often investigated in addition to the EA.

It is known that sEMG may provide objective means to assess sports seizures, aid the diagnosis process, and may help improve treatment outcomes [7]. Therefore, EMG, together with IMU data, has been proposed to monitor the progress of diseases linked to a dysfunctional nervous system such as multiple sclerosis and which may lead to limb dysfunction [8].

EMG has been a popular sensor in the detection of ataxia-type disorders, for instance, in the analysis of cerebellar, mitochondrial, and spinocerebellar ataxia [9]–[11], especially focusing on tremors caused by the disease [12] movement disorders in targeted exercises [13], as well as for early detection of the disease in risk groups [14].

We hope that, by using EMG and IMU sensory systems, indicators for the MIRAS disease and its progression may be identified for the patients and that the technology would make it possible that eventually, patients may become capable of performing tests and analysis on their own, using a mobile sensor system, so that the monitoring of the progression of the disease may become less subjective and involving significantly more measurement points and thus more frequent feedback on the success of treatment and on the progress of the disease.

III. STUDY DESCRIPTION

Accompanied by a medical expert, we have conducted a study comprising eight volunteers (cf. table I). The participant-ID in the 'ID' column comprises a random letter identifying the participant, followed by an indicator specifying whether the left ('L') or right ('R') hand has been used during the measurements. The MIRAS patient has the ID 'M' in the last column. All participants have been right-handed, and while a variation of body conditions, age, and gender have been considered, similarity with the patient is in particular interesting from these statistics. For subjects A and B, we conducted the tasks with both hands for comparison. The patient had suffered from MIRAS for 28 years. Since the high BMI of the patient (M) might impact the sEMG measurements, a matched control participant (E) with a similar physique was included.

We used two Delsys Trigno Quattro EMG sensors, which feature four bipolar dry sEMG electrodes (10mm inter-electrode distance) and an IMU with accelerometer, gyroscope, and magnetometer. The sensors were connected to the Delsys Charge-4 Station, and data was recorded via the EMG Logger smartphone application installed on an android smartphone.

During the measurement, four electrodes were placed according to the SENIAM guidelines [15]3 on the long and lateral heads of the triceps brachii muscle, biceps brachii, and brachioradialis to record the elbow flexion and extension activities. We sampled raw sEMG at 1111Hz at a range of +/- 5 mV (Mode 'QC 12.5 EMG 16 Bit') while the second sensor was placed on the back of the hand to collect IMU data from there. All data were converted to .xlsx files through the EMG Logger app, and further processing was conducted using python.

In designing tasks, we focused on larger arm movements since MIRAS patients are frequently wheelchair-bound and experience difficulties with precision tasks. These tasks included measurements with a relaxed arm, maximal efforts, static contractions, exhaustion tasks, a pointing task (nose-finger test). For the data from participants A and B, some tasks were performed slightly differently and without IMU.

A. Pre-processing

Due to external factors (drop in Bluetooth connection; app instability), we had to clean the data from blank (zero values) measurement rows. Further, in some signals, spikes of large amplitude sporadically occurred in the sEMG data due to tension of cables while performing tasks. We addressed this by disregarding values that exceeded the 95th percentile of the signal more than 2.5 times. Finally, noise was removed using a 4th order Butterworth band-pass filter (cut-off at 20 Hz and 450 Hz), which is the same as the pre-processing performed by the device itself. Where necessary, powerline interference at 50 Hz and its harmonics were removed using 2nd order IIR Notch filters. For the computation of the EA using the root-mean-square (RMS) and the MF window sizes of 250ms were used based on recommendations in [16], [17].

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3https://delsys.com/trigno-quattro

TABLE I

<table>
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<th>Gender</th>
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<th>Height (m)</th>
<th>BMI (kg/m2)</th>
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<td>40.7</td>
</tr>
</tbody>
</table>

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2https://delsys.com/trigno-quattro
3http://seniam.org/
B. Normalization

The absolute EA [Volt] of the measured data strongly depends on the individual so that absolute EA values are not directly comparable across participants. For instance, due to the high BMI, the patient (M) and the matched control (E) stand out with especially low EA levels. Therefore, we computed for each muscle the relative EA (%) by subtracting its baseline EA before normalizing to the maximal EA. For each participant separately, we determined the baseline EA as the lowest of the 25th percentile over multiple recordings of keeping the arm completely relaxed (relaxed arm task). In addition, maximal voluntary contraction (MVC) for elbow flexion and extension were measured to capture the maximal EA of the muscle (maximal efforts task).

IV. RESULTS

We will now discuss observations from the measurements.

A. Static tasks

We instructed the participants to hold weights in position both for elbow flexor muscles and for the triceps (figure 1). Generally, a higher weight requires a higher EA, and the extensor muscle showed higher EA levels for the same weight. Due to the general muscle weakness, ataxia, as well as lack of training, the patient stands out with the highest EA levels.

To estimate the unsteadiness in the muscle activity, we calculated the standard deviation of the EA (figure 2). While the absolute standard deviation is exceptionally high for the patient, this is attributed to the patient’s higher EA level as visible after normalization (right figure). Concluding, at the level of individual capability, the patient’s muscles did not vary more than others.

B. Exhaustion tasks

Measurements were concluded with static exhaustion tasks for elbow flexor and extensor muscles. The tasks were identical to the static tasks but with a 2 kg weight held for as long as possible. Commonly, an increase of the EA and a decrease of the sEMG MF are associated with muscular fatigue. We were expecting that the muscular deficits of the patient would stand out. Since MIRAS affects the mitochondria, which produces muscle energy, their deficits might be more visible.

Indeed, holding the weight became too difficult for the patient within 40 s (flexor task) and 20 s (extensor task), while other male subjects were not exhausted after more than 2 minutes, after which the measurement was stopped.

For the patient, an EA increase could be observed for 3 out of 4 muscles during their respective task, and an MF also decreases for 3 muscles. For other subjects, EA increase and MF decrease were both observable in 70% (jointly in 50%) of the measures. In terms of the magnitude, the patient does not stand out particularly.

Overall, a certain relative weakness of the patient could be seen in the higher muscular activity, the shorter fatigue time, and the difficulties the patient had during the tasks with weights. This weakness can at least partially be attributed to the consequences of MIRAS, but also the relative weakness of the patient (cf. static tasks) may explain these differences.

We note that it is challenging to analyze fatigue in this way since EA and MF changes may also be caused by factors such as muscular activity, which may be affected by subconscious compensation strategies. For follow-up studies, we suggest quantifying patients’ strength by measuring the maximal force (torque) for flexors and extensors with a load cell during the MVCs in order to perform the static tasks at the same relative strength (e.g., at 20% of the maximum strength of participants), which could yield in a better comparison.

C. Ataxia indicators from the pointing task

We performed variations of the finger-nose test. While all participants were instructed to perform the task at their own speed, the patient was slowest with approximately 2.9 seconds pointing from nose to the target and back (other subjects: average: 2.46s, slowest: 2.78s). The patient also stands out regarding the average magnitude of the EA (figure 3). The biceps showed high muscle activity for almost all subjects. The high EA levels for the patient are probably attributed to the limited strength of the patient. In the following, we discuss various indicators for ataxia, extracted from the EAs while performing the pointing task.

\[^4\text{No EA increase observable for the biceps. No MF decrease is observable for the lateral head of the triceps.}\]
1) **Co-contraction:** Following the results by [20], we expected that ataxia would result in high co-contraction (activation of opposing muscle groups) during the pointing task, while healthy subjects would show high co-activation levels (coordinated muscle behavior). Interestingly, these movement patterns were also person-dependent for healthy subjects, and we found both co-activation (figure 4, left) and co-contraction (right) patterns in healthy subjects.

Figure 5 plots the correlation of the EA signals for the arm muscles [20]. A high positive correlation of two muscles indicates simultaneous activation. This can be seen for the two heads of the triceps for all subjects. A negative correlation indicates that the muscles are not active at the same time. While plausible for antagonistic muscles (e.g., biceps and triceps), we observed very different values for the antagonist correlations. Interestingly, the matched control stands out most with a positive correlation of biceps and triceps and a negative correlation of the brachioradialis to all other muscles. Overall, inter-individual differences seem apparent in the pointing task.

2) **Frequency of EA components:** To evaluate the instability and amount of random muscle activity, the standard deviation of the EA signal could be an appropriate measure. For this, we separate the movement from unnecessary muscle activity, and we filter the signal with a cut-off at 1Hz (assuming task execution frequencies of 0.35 Hz to 0.45 Hz for the pointing task). Lower frequency components might be attributable to the movement, while others are more likely due to random or unnecessary muscular activity. Figure 6 shows that indeed the low-frequency components dominate the standard deviation for all muscles and subjects. A comparatively high ratio can be observed for the patient, so this might indeed be a useful measure to identify ataxia.

The median frequency of the EA is shown in Figure 7, which is not to be confused with the MF of the raw sEMG signal. Again, the patient stands out. Concluding, despite the slower task execution (lower frequency of the main movement), the patients’ EA contains more high-frequency components.

3) **Similarity of repetitions:** We observed apparent intra-individual differences of different repetitions of the pointing task both for the healthy subjects (Figure 4) and for the matched control and the patient (Figure 8). These differences are observable in the EA of a muscle during the task and
the timing of the muscle activation. They can be attributed to natural random variations in the sEMG signal and slight variations in the task execution.

Compared to the control subject (E), the patient showed more irregularities in the activation. To quantify this, we compute the correlation of one muscle’s EA during repetitions of the task. A high correlation then indicates almost identical task executions, while a low correlation indicates variation. The patient reached the lowest correlation during repetitions for 2 muscles and also had the lowest self-correlation measures in general (Figure 9). However, individual differences complicate the evaluation of ataxia using this feature. The lowest self-correlation was achieved by subject D (brachioradialis), since this subject barely used the muscle at all during this task (EA below 0.5% of the maximum) (Figure 4). Self-correlation might be useful in quantifying the difficulties a patient would have repeating a task identically, possibly a result of ataxia.

D. Variations and alternative tasks

We performed two variations of the pointing task, namely holding a 0.5 kg weight during the task (resulting in higher EA levels) and stopping between repetitions for one second at both nose and target. While holding a weight, differences between patients and control to the normal execution were less pronounced concerning the extent of high-frequency components of the EA but more explicit concerning the muscles’ self-correlation. For the interrupted movement, observations were very similar to the ordinary pointing task. Overall, the different versions did not add much to the current analysis but could be useful when further investigating more specific aspects.

Additionally, the patient and the controls performed some other tasks, such as alternating with an object between two points on the table. We realized that the problem of inter-individual differences seemed more pronounced in this case as participants tended to execute the task using shoulder joints. Furthermore, hand rotation (pronation and supination), as well as wrist movement (extension, flexion, and radial and ulnar deviation), were measured. We recorded sEMG of some involved muscles but without appropriate maximal contractions for each muscle. The patient had more difficulties during the tasks than the controls, and the sEMG signal appears to be less regular. Due to the bigger number of muscles involved, attributing differences to the ataxia might require a more task-specific analysis.

E. Analysis of IMU signal

In addition to the sEMG, we collected IMU data from the back of the active hand during task execution. The IMU has the advantage that normalization to a specific task is not required and contains less noise.

For instance, the IMU may indicate the steadiness of the static tasks through acceleration (Figure 10). We observed that the hand of the patient was less stable. Possibly, this reflects the lack of coordination, which may be caused by (sensory) neuropathy of the patient due to MIRAS. While a certain level of variation of the EA is entirely normal, and a healthy person may be capable of compensating this by using other muscles and joints, the patient does not seem capable of doing so.

The features found for the EA signal also seem to be present in the IMU signal with more variation of high-frequency components and less similarity in the task executions. These observations in the IMU signal can probably be traced back to the lack of coordination of the patient’s muscles visible in the EA signal. The movement difficulties caused by MIRAS seem to be passed on from the muscle to the hand and the IMU. The IMU signal could be a simple summary of the coordination difficulties of the muscles.
V. CONCLUSION

Overall, the results of this study indicate that sEMG recordings of the arm can be used to identify and quantify features related to the ataxia of a MIRAS patient. However, some confounding factors also have to be considered.

The patient’s relatively high amount of subcutaneous fat in the arm can be addressed with normalization of the EA and an adequate control group. Many differences observed between the patient and controls may be traced back to the patient’s relative weakness, which in turn is attributable to the ataxia. It would be complicated to distinguish effects caused by this weakness from effects of the ataxia itself as they go hand in hand. As seen for the pointing task, inter-individual differences in muscle usage can make the analysis more complex, especially for movements. Therefore tasks should be clearly defined as possible to reduce differences in the task execution. For tracking the disease progression in one patient, the inter-individual differences may be less relevant.

From the present analysis, it is not clear if MIRAS causes changes in how the muscle is activated. Such differences might be better identifiable using the raw sEMG signal. Here, Machine Learning may be of advantage in handling the relatively enormous amount of data. On a larger scale, however, differences of patient and controls were found looking at the EA. The patient showed a higher amount of unnecessary muscular activity and less regularity in doing the pointing task. Both could possibly be traced back to coordination difficulties caused by MIRAS. Some intra-individual differences were also observed for healthy patients, but they seem to compensate for varying muscular activity levels. These mechanisms would need to be further investigated since it possibly means that the ataxia is more apparent in the resulting movement, which can be observed with an IMU.

The current project has shown some possibilities but also limitations of analyzing the ataxia resulting from MIRAS using sEMG. Based on this, some additional aspects, such as positional information from the IMU, could be investigated in depth before advancing to more measurements. Experiments with patients and matched controls could be performed and repeated after six months to indicate the usefulness of features for tracking MIRAS progression.

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