Enhancing clinical assessment in children with neuromuscular diseases with kinematic parameterization: a one-year follow-up

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Introduction

Duchenne muscular dystrophy (DMD) and spinal muscular atrophy (SMA) are rare genetic disorders characterized by progressive degeneration and weakening of muscles[1]. Traditionally, protocols based on clinical scales have been used to assess the motor function of children with such disorders. Although these scales provide a standardized approach for evaluating motor function, they rely on the clinician's subjective interpretation, which can introduce variability into the assessment[2]. To address this limitation, instrumented analysis, particularly with Inertial Measurement Units (IMUs), has gained popularity, providing objective data to complement specialists' assessments.

This study aims to assess the effectiveness of IMU-based metrics in tracking the upper limb motor function of non-ambulant children over time to capture the evolution of the disease and provide insights into the rate of motor function decline.

Material & Methods

Four children with DMD (11-14 years, 1-3 Brooke score) and five with SMA (11-15 years, 2-3 Brooke score) have been analyzed while performing the clinical scales. The study was approved by the Ethical Committee of the Hospital Sant Joan de Déu (HSJD), Barcelona. The assessments were conducted at baseline and then repeated one year later on the same participants.

The data of seven IMUs (Xsens Dot, Xsens Technologies), placed on the upper limb were recorded while performing the scales. Each IMU provided quaternions data which were used to compute the kinematics of a seven-segment upper body model, according to the standards[3]. The calibration was conducted with the subject seated on the wheelchair with forearms laying on a table. Pictures of the patients in the frontal and sagittal planes were taken during the calibration process. To improve calibration accuracy, the information of the pictures was integrated with IMUs data[4]. Range of motion (ROM) of the shoulder and workspace area (A) of the hand, in the frontal and horizontal planes, were evaluated as clinically relevant metrics[5]. The workspace area was normalized to the maximum area achievable by the child (in %).

Results and Discussion

Fig. 1 shows the workspace area in the frontal plane for children with DMD, according to Brooke score. The dots on the left represent the area reached during the first assessment, while those on the right represent the area reached during the follow-up assessment, one year later. Despite a general decrease in the frontal area over time, one child shows an increase, which aligns with an improved Brooke score, indicating a distinct motor function development pattern. Similar results were found for ROM of the shoulder and horizontal workspace area. The kinematic data for children with SMA show more variability, indicating the need to analyze more cases.

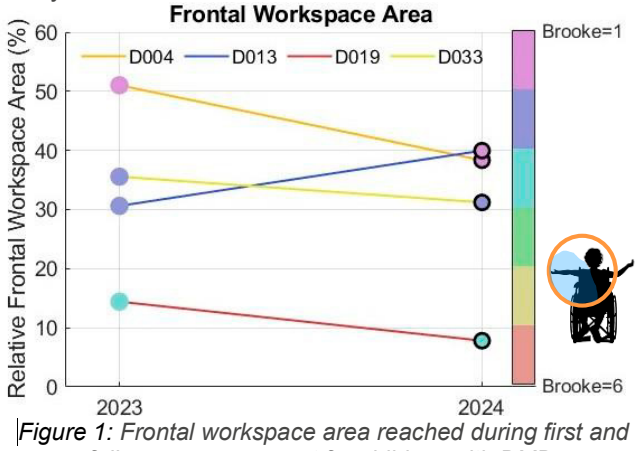

Figure 1: Frontal workspace area reached during first and follow up assessment for children with DMD.

Conclusion

This work represents a first step toward the use of an IMU-based system to evaluate the progression of neuromuscular diseases across different stages. These preliminary results suggest that some kinematic metrics may have potential to objectively track disease progression over time. Future research should focus on determining whether an IMU-based system can detect subtle changes in motor function that may not be captured by conventional clinical assessment, analyzing a wider group of patients.

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