Digital Approaches for Myositis

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Abstract

Purpose of Review This article serves as a comprehensive review, focusing on digital approaches utilized in the diagnosis, monitoring, and treatment of patients with idiopathic inflammatory myopathies (IIM). The authors critically assess the literature published in the last three years, evaluating the advancements and progress achieved in this specific domain.

Recent Findings Remarkable strides have been made in the realm of digital diagnostic support, particularly in image analysis and clinical prediction models, showing promise in aiding the diagnosis of IIM. The field of remote patient monitoring has also witnessed significant advancements, revolutionizing the care process by offering more convenient, data-driven, and continuous monitoring for IIM patients. Various digital tools, such as wearables, video- and voice consultations, and electronic patient-reported outcomes, have been extensively explored and implemented to enhance patient care. Survey studies consistently reveal a high acceptance of telehealth services among patients. Additionally, internet-based studies have facilitated the efficient and rapid recruitment of IIM patients for research purposes. Moreover, the integration of sensors and exoskeletons has shown great potential in significantly improving the functionality and quality of life for individuals with muscle weakness caused by IIM.

Summary The integration of digital health solutions in the care of IIM patients is steadily gaining attention and exploration. Although the existing evidence is limited, it does indicate that patients can be adequately and safely supported through digital means throughout their entire healthcare journey. The growing interest in digital health technologies holds the promise of improving the overall management and outcomes for individuals with idiopathic inflammatory myopathies.

Keywords Myositis · Telehealth · eHealth · Wearables · Digital

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Introduction

Idiopathic inflammatory myopathies (IIM) encompass a group of chronic and rare autoimmune disorders primarily affecting the proximal muscles. The complexity, non-specific symptoms, and rarity of these diseases contribute to a significant diagnostic delay [1]. Disease activity tends to fluctuate, with intermittent flares between medical visits, necessitating timely adjustments to immunosuppressive treatments. Current monitoring methods relying on face-to-face (F2F) visits pose significant burdens on patients with limited mobility and only provide a snapshot for healthcare professionals (HCPs). Access to specialized centres for IIM patients is limited and often challenging for patients to reach [2]. Treatment approaches for IIM patients remain suboptimal, often involving a trial-and-error process, and the provision of prescribed physiotherapy is lacking [1].

Technological advancements continue to revolutionize the healthcare landscape, offering substantial potential to

enhance IIM healthcare significantly. Embracing a digitally supported patient pathway holds the promise of providing more patient-centric, convenient, cost-effective, continuous, and data-driven care. The COVID-19 pandemic has expedited the adoption of telehealth solutions, leading to increased acceptance among patients and rheumatologists [3, 4]. The benefits of telemedicine have been acknowledged by EULAR, as evidenced by their publication of points-to-consider for remote care in 2022 [5••]. These developments underscore the transformative role that digital health solutions can play in optimising the management and outcomes for individuals with inflammatory myopathies [6].

By critically evaluating the recent literature, we aim to provide a comprehensive understanding of the current state of digital health solutions for IIM and identify potential avenues for future research and implementation in clinical practice.

Diagnosis

Digital health tools, particularly patient-facing diagnostic decision support systems (DDSS), have the potential to expedite the diagnosis of IIM patients. Rheumatic patients are increasingly utilizing such tools [4, 7], and they have been subject to investigation in a large multicentre trial for individuals with suspected rheumatic diseases [7–9]. The key advantage of these apps, also known as symptom checkers, lies in their accessibility and ability to provide a rapid initial assessment.

While patients have shown favourable perceptions towards DDSS, their diagnostic accuracy remains relatively low. This could be attributed to the limited information currently incorporated into these tools, primarily reliant on the presence or absence of symptoms. Enhanced access to a broader range of information is likely to improve diagnostic accuracy, as demonstrated in other studies, such as asynchronous telehealth assessments in patients with suspected axial spondyloarthritis [10]. Expanding the depth and breadth of information available to these digital tools could potentially enhance their efficacy in aiding the diagnosis of inflammatory myopathies and other rheumatic conditions. Determination of autoantibodies remains a diagnostic gold standard to differentiate the heterogenous forms of IIM. A noteworthy study demonstrated that patients with IIM can effectively collect capillary blood samples at home, and the analysis of these samples proves to be as accurate as venous blood collected in a hospital setting [11]. Patients showed great enthusiasm in avoiding unnecessary appointments solely for blood collection purposes, particularly when utilizing new upper-arm devices that were found to be almost painless [12]. At-home diagnostic assessment combining eHealth and

patient self-sampling could significantly improve access to care and accelerate diagnosis.

Other digital medical devices enable earlier detection of relevant complications by augmenting physicians' ability in the diagnostic process. Xue et al. built a machine learning algorithm, superior to a conventional regression model to predict the risk of antiMDA5 antibody in juvenile dermatomyositis children [13]; a fatal manifestation of IIMs is interstitial lung disease (ILD). Early identification, monitoring of the lung areas involved and correct classification of ILD pattern is crucial. Artificial intelligence is especially implemented in medical image analysis. A systematic review pointed out diagnostic accuracies of 78%–91%, with two studies demonstrating near-expert performance. However, most studies (78.9%; 15/19) had a high risk of bias.

Monitoring

A recent international survey among rheumatologists showed that the majority of participants felt that telemedicine is inadequate for the monitoring of IIM flares, compared to flares of arthritis [14]. Disease complexity, lack of financial reimbursement and lack of physical examination were main identified barriers, in line with previous work [15, 16]. Nevertheless, during the COVID pandemic remote monitoring for IIM patients was implemented at some centres. Cavagna et al. presented results of their telehealth implementation during the pandemic for patients with connective tissue disease, including IIMs [17•]. Confirming previous results [18], patients' attitude towards telehealth correlated with the distance from the hospital. Patients with a higher education were more likely to prefer telemedicine over in-person visits. Lack of telemedicine skills was a main barrier, highlighting eHealth literacy [19] as major factor for successful implementation of telehealth. Naveen et al. also offered IIM patients' telehealth during the pandemic [20•]. Voice consultations were the preferred medium and nearly one third of the patients consulted in emergency cases. In this study, the authors did not observe differences between telehealth users and non-users regarding their demographic and socioeconomic profile.

Most of the recent digital health publications regarding IIMs concern disease monitoring. The clinical gold-standard are collected during F2F visits, enabling only a limited analysis of disease activity. Digital health, including wearables, videoconsultations, mobile apps and ePRO (electronic patient-reported outcomes) enable a more continuous monitoring of disease activity, complementing F2F visits. The current gold-standard to assess muscle strength included as a core set measure (CSM) in the official ACR/EULAR total improvement score (TIS) [21] is manual muscle testing (MMT-8). The test remains the gold-standard despite being very subjective,

subject to large interobserver variability and exhibiting ceiling effects [22]. Focusing on muscle endurance instead of direct muscle strength testing tries to overcome these barriers. The clinical gold standards to measure muscle endurance are the functional index (FI) [23] and 6-min walking distance (6MWD). The latest and third version of the FI includes three muscle groups and takes less than 10 min: shoulder flexion, neck flexion and hip flexion. One disadvantage is that patients are required to complete the repetitions with a specific weight and at a set rhythm. Additionally, the FI and 6MWD are performed in the hospital under HCP supervision. Patient-performed tasks and device-based measurements enable remote monitoring of disease activity.

Validation of remote assessments with IMACS core set outcomes is the unmet need of the hour. Ravichandran N et al. recently evaluated two novel patient–performed tests, namely ten times arm lift (AL) test and 2-min walk distance (2MWD) in IIM patients in a telehealth myositis patient cohort [24•]. AL correlated well with all CSMs except CK levels and MDI. 2MWD values were highly variable and showed no correlation with CSMs.

Wearables have the advantage of continuous and passive data collection. Rockette-Wagner et al. validated a waist-worn sensor (ActiGraph GT3X-BT) [25]. Measurements showed good TIS responsiveness. The same group compared the accuracy of the expensive ActiGraph® with the more affordable Fitbit® devices and could demonstrate good agreement and strong correlation (ICC 0.96) for step count [26••]. Milazzo et al. published a pilot study with wearable sensors to objectively quantify muscle strength of the upper limb muscles in patients with muscle dystrophies [27]. Whereas the current system is still quite bulky, a point-of-care solution for at home monitoring seems feasible. Using data collected passively from smartwatches and/or smartphones, a self-administered 6-min walk distance is very convenient for patients [28].

Established questionnaires can easily be distributed electronically. Implementation of a smartphone app and a weekly questionnaire unveiled the high frequency of disease flares in IIM patients [29]. The most commonly used questionnaires in IIM are Health Assessment Questionnaire (HAQ-DI) and SF-36 physical function-10 (PF10). Recently Yoshida et al. established the dispersion of patient reported physical function in IIM [30], and a global patient led e-survey identified the spectrum of pain [31] experienced by patients using validated PROs.

Treatment

Effective remote monitoring could provide a more solid and detailed rationale for treatment decisions. Despite the recent advances in drug treatment of IIMs [32, 33], some patients experience are very refractory and can no longer manage their daily lives independently. Wearables such as a cyborg Hybrid Assistive Limb investigated by Nakajima et al. [34] could help patients to cope better with everyday life. In a multicentre randomized controlled trial in patients with neuromuscular diseases patients using the wearable device experienced significant improvements regarding 2MWD and MMT scores. For the Mysosuit, a robotic exoskeleton-assisted wearable, first promising results have also been reported in a pilot study with lung transplantation candidates [35].

Internet-based research is invaluable for rare diseases such as IIM to accelerate research [36]. A recent large international survey study in patients with antisynthetase syndrome enabled a detailed picture of healthcare utilization and unmet patient needs [1]. The study unveiled a general lack of prescribed physiotherapy and heterogenous treatment patterns. Patients were most interested in additional disease related information and preferred online information compared to local community workshops. Joshi et al. systematically identified and evaluated the quality of YouTube videos for patient and physician education on inflammatory myositis [37]. A total of 900 videos were identified and 74% of the videos were classified as useful. The authors conclude that HCPs should recommend and provide their patients with high-quality information material. A recent app-based pilot study prompting patients' evidence-based information suggested an improvement of quality of life for arthritis patients [38] and offer for IIM patients would likely just be as useful.

Research

Social media driven internet-based research allows greater outreach and data collection for rare rheumatic diseases such as IIM. This is best exemplified by the COVAD group, a focussed patient reported e-survey floated by a collaborative group of experts covering 106 countries and 164 centres [39]. This group identified global disparities in treatment of IIM, outlining advocacy for rare rheumatic diseases as an important agenda for collaborative study groups with worldwide representation [40]. Hannah et al. demonstrated that based on routinely collected clinical data, namely ICD codes, IIM patients can be accurately identified to enable more cost-effective research [41]. A combination of ICD codes improved diagnostic accuracy, paving the way to optimise the use of registry-based datasets for rare disease research.

Conclusion

Given the limited availability of specialized centres and the severe functional limitations caused by IIM, telehealth emerges as an ideal solution for improving patient care.

This review emphasizes the substantial progress achieved in recent years, although large and robust studies in this area are still relatively scarce. The majority of identified studies have focused on remote patient monitoring, indicating a growing interest in this approach. Both patients and healthcare professionals express a strong desire for a more detailed and continuous overview of disease activity between face-to-face appointments. The integration of ePROs, self-collection of blood samples, and wearables empowers patients to take on a more active role in managing their condition.

Notably, the implementation of telehealth could also pave the way for a real-world data registry, facilitating patient recruitment, and even enabling decentralized clinical trials. These advancements have the potential to significantly enhance the overall management and research related to idiopathic inflammatory myopathies, offering greater convenience and improved outcomes for patients.

Data Availability The data that support the findings of this study are available from the corresponding author, Johannes Knitza, upon reasonable request.

Compliance with Ethical Standards

Ethical Approval Not required.

Conflict of Interest The authors declare no competing interests.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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