Poor Quality of Life and Substantial Disability in Adult Patients with Dermatomyositis and Polymyositis

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Introduction

- Dermatomyositis (DM) and polymyositis (PM) are rare autoimmune inflammatory myopathies characterized by muscle weakness and multiple extra-muscular manifestations that have a detrimental impact on patients' lives. The current evidence focuses on the clinical outcome of the disease, and little is known about the true impact of DM and PM on patient's health-related quality of life (HRQoL). The goal of our study was to systematically review and summarize evidence on humanistic burden of disease in adults with DM and PM.

Methods

- A systematic literature review (SLR) was conducted in MEDLINE and Embase to identify studies in children and adults with DM and PM, published in English between Jan 1, 2011, and Apr 28, 2021. Studies enrolling at least 10 patients were included, irrespective of country or region. The current poster summarizes data on humanistic burden of disease in adults with DM and PM.

Results

- The SLR yielded 222 studies described in 229 publications of which 16 studies[1-16] reported data on HRQoL, daily functioning or disability-related outcomes in adults with DM and PM (**Figure 1**), The majority of studies were conducted in the US and EU (**Figure 2**).

Figure 1. PRISMA diagram

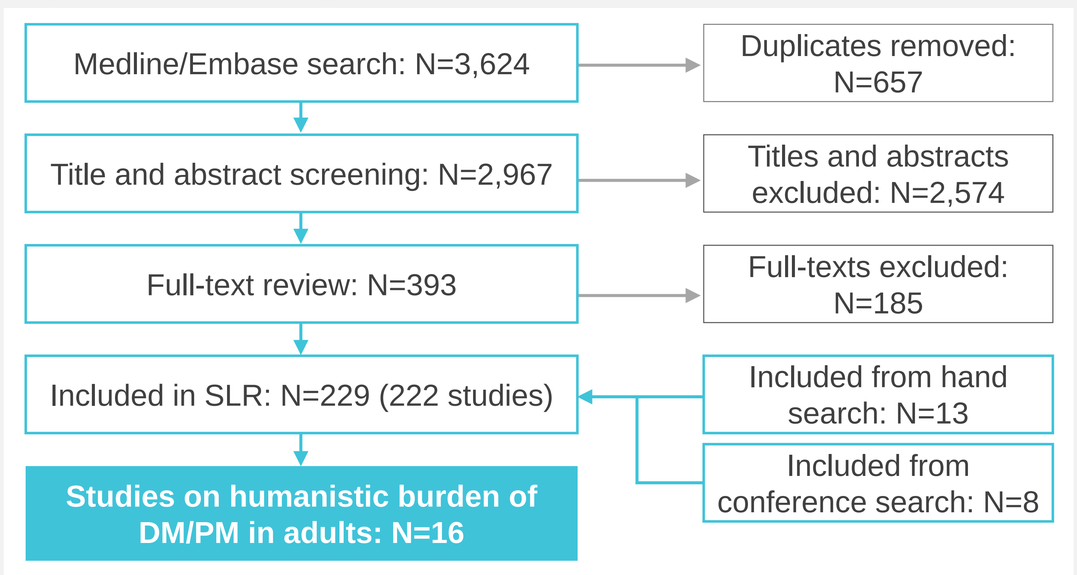
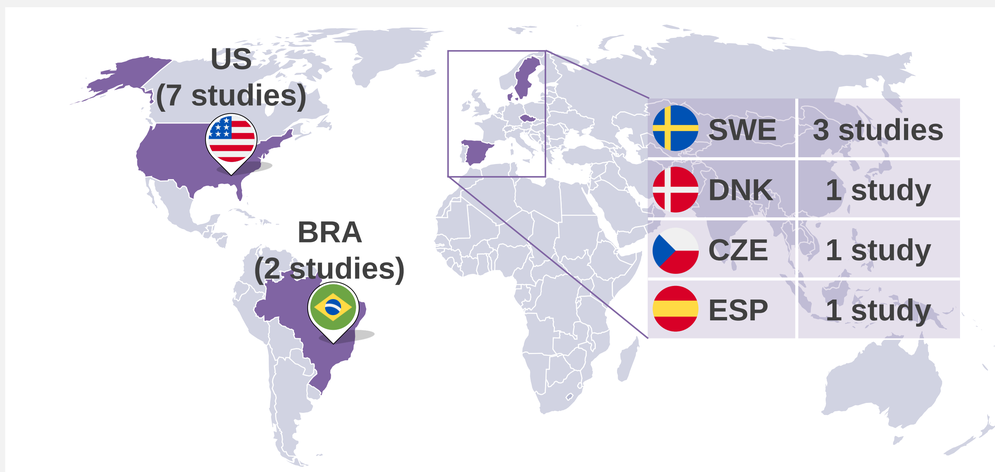


Figure 2. The majority of studies reporting humanistic burden of DM and PM in adults were conducted in the US and EU



* One study was conducted across multiple continents, not included in the map.

- DM and PM had detrimental impact on patients' physical, psychosocial and social role functioning. DM and PM patients reported significantly worse HRQoL across multiple SF-36 domains than matched general population[3,5,6,11] and patients with non-skin conditions[3] (**Table 1**). Moreover, DM adults had significantly worse HRQoL measured by Skindex-29 than multiple other skin disorders, such as cutaneous T-cell lymphoma, rosacea, and vitiligo.[3]
- Multiple studies showed disability and progressive loss of independence due to DM and PM. In a Swedish cohort study, muscle strength measured by MMT8 was as an independent factor for poor HRQoL in the physical domain of the SF-36 (p=0.006).[11]

Results (cont'd)

Table 1. DM and PM adults had significantly worse HRQoL in multiple SF-36 domains compared to non-skin conditions and general population

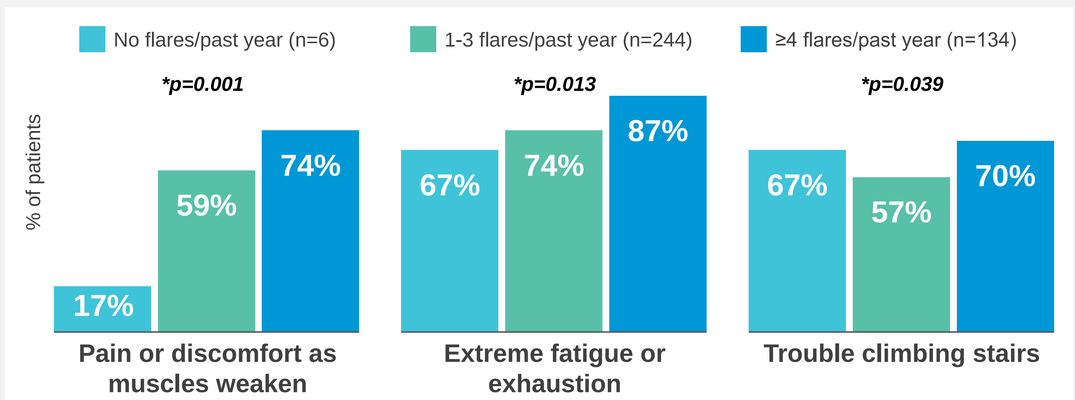
	Study cohort	Control cohort	DM/PM vs Control
🇺🇸	**DM (N=120)[3]**	HTN (N=2,089)	**Worse scores** in all SF-36 domains except BP (p≤0.006)
🇺🇸	**DM (N=120)[3]**	T2DM (N=541)	**Worse scores** in SF-36 VT, SF, MH domains (p≤0.001)
🇺🇸	**DM (N=120)[3]**	Recent myocardial infarction (N=107)	**Worse scores** in SF-36 VT and SF domains (p=0.001)
🇺🇸	**DM (N=120)[3]**	Clinical depression (N=502)	**Worse scores** in SF-36 PF domain (p=0.001)
🇺🇸	**DM (N=120)[3]**	CHF (N=216)	**Worse score** in SF-36 MH domain (p=0.003)
🇺🇸	**DM (N=120)[3]**	Gen. pop. (N=2,474)	**Worse scores** in all SF-36 domains (p<0.05)
🇸🇪	**DM/PM (N=31)[5]**	Age-gender matched gen. pop. (N=NR)	**Worse scores** in all SF-36 domains (p<0.05)
🇸🇪	**DM/PM (N=48)[6]**	Gen. pop. (N=NR)	**Worse scores** in SF-36 PF, RF, GH, VT, SF domains (p≤0.02)
🇩🇰	**DM/PM (N=75)[11]**	Age-gender matched gen. pop. (N=48)	**Worse scores** in SF-36 PCS and MCS (p<0.001)

In a Danish study, patients with DM and PM showed reduced grip force vs age-gender matched general population that significantly impaired their ability to perform domestic activities measured by Myositis Activity Profile (MAP) (p<0.05).[5] Median MAP scores ranged from 2 to 6 points suggesting slight to very high difficulties in performing daily activities with work and leisure time activity being impaired the most.[5]

In an international survey among 183 patients with DM and PM, 27-48% of respondents depended on caregivers for various daily activities and and up to 38% of them required walking aids or other facilitating devices. Overall, 40% and 65% of DM and PM patients reported moderate-to-very severe disability in the Health Assessment Questionnaire (HAQ).[12]

In the US, patients with DM more frequently reported clinically significant fatigue (SF-36 vitality score ≤ 35) than heathy controls, 32% vs 2%, respectively; p<0.01).[15] Extreme fatigue/exhaustion, pain and trouble climbing stairs were frequently reported by DM and PM patients and showed increased tendency with disease flares (**Figure 3**).[1]

Figure 3. Pain as muscles weaken, fatigue/exhaustion and trouble climbing stairs significantly increased with frequency of DM/PM flares



Conclusions

- **Patients with DM and PM suffer from a high level of disability and poor HRQoL across multiple domains of life, including the physical, psychological, and social role functioning. The burden of disease is similar between US and European patient populations.**
- **There is a high unmet need for a therapy that can improve HRQoL in patients with DM and PM.**

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Abbreviations: **BP**, bodily pain; **CHF**, congestive heart failure; **DM**, dermatomyositis; **EU**, European Union; **GH**, general health; **HAQ**, Health Assessment Questionnaire; **HRQoL**, health-related quality of life; **HTN**, hypertension; **MAP**, Myositis Activity Profile; **MCS**, Mental Component Summary; **MH**, mental health; **NR**, not reported; **PCS**, Physical Component Summary; **PF**, physical functioning; **PM**, polymyositis; **RF**, role functioning; **SF**, social functioning; **SF-36**, Short Form 36 Health Survey Questionnaire; **SLR**, systematic literature review; **T2DM**, Type 2 Diabetes Mellitus; **VT**, vitality

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