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HUMANISTIC BURDEN OF HUNTINGTON'S DISEASE (HD): A SYSTEMATIC REVIEW

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Background

- Huntington's disease (HD) is a rare, progressive, genetic neurodegenerative condition characterized by cognitive, motor, and psychiatric dysfunction that affects about 43,000 people in the US and 40-100 per million people in Europe.
- The HD prevalence was reported to be 15.2 per 100,000 for Medicaid beneficiaries under 65 years of age[1]
- A substantial increase in HD prevalence was reported in the UK, from 4.3 per 100,000 persons in 2000 to 13 per 100,000 in 2018

Objective

- The systematic review aimed to identify and summarize the health related quality of life (HRQoL) and patient reported outcomes (PROs) in HD

Methodology

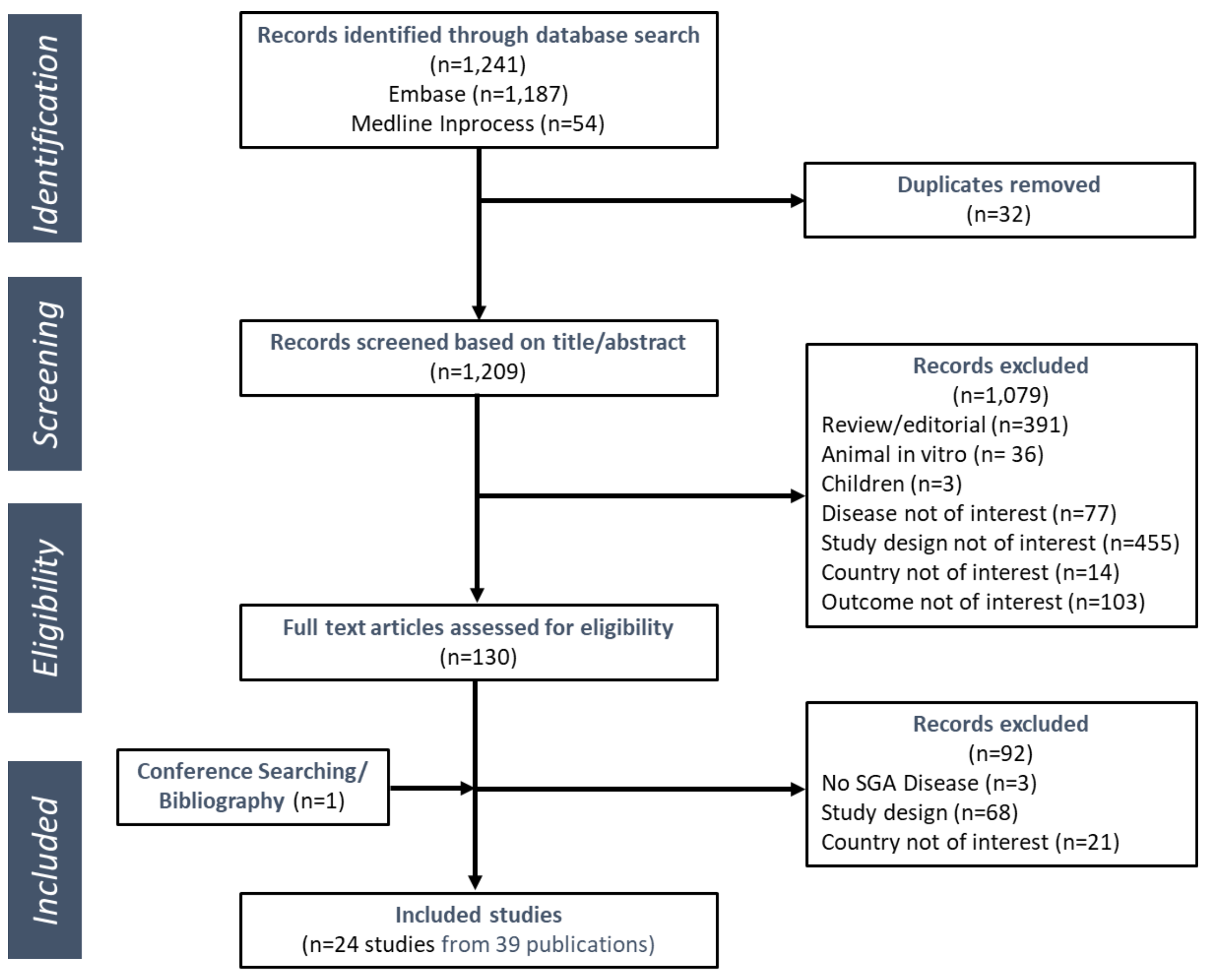
- Key biomedical databases (Embase, MEDLINE, MEDLINE in process) were searched from database inception to May 2022
- The search terms included different combinations of HD along with quality-of-life measures
- Two independent reviewers performed the screening and data extraction activities with conflicts resolved by a third independent reviewer

Table 1: Study inclusion criteria

PICOS	Inclusion
Population	Adult patients with HD
Intervention	No restriction
Comparator	No restriction
Outcome	HRQol, PROs
Study Design	Cohort studies Cross-sectional studies Observational studies Surveys
Geography	US , EU4 and UK

Results

Figure 1: PRISMA Diagram



Records identified through database search
(n=1,241)
Embase (n=1,187)
Medline Inprocess (n=54)

Duplicates removed
(n=32)

Records screened based on title/abstract
(n=1,209)

Records excluded
(n=1,079)
Review/editorial (n=391)
Animal in vitro (n= 36)
Children (n=3)
Disease not of interest (n=77)
Study design not of interest (n=455)
Country not of interest (n=14)
Outcome not of interest (n=103)

Full text articles assessed for eligibility
(n=130)

Conference Searching/ Bibliography (n=1)

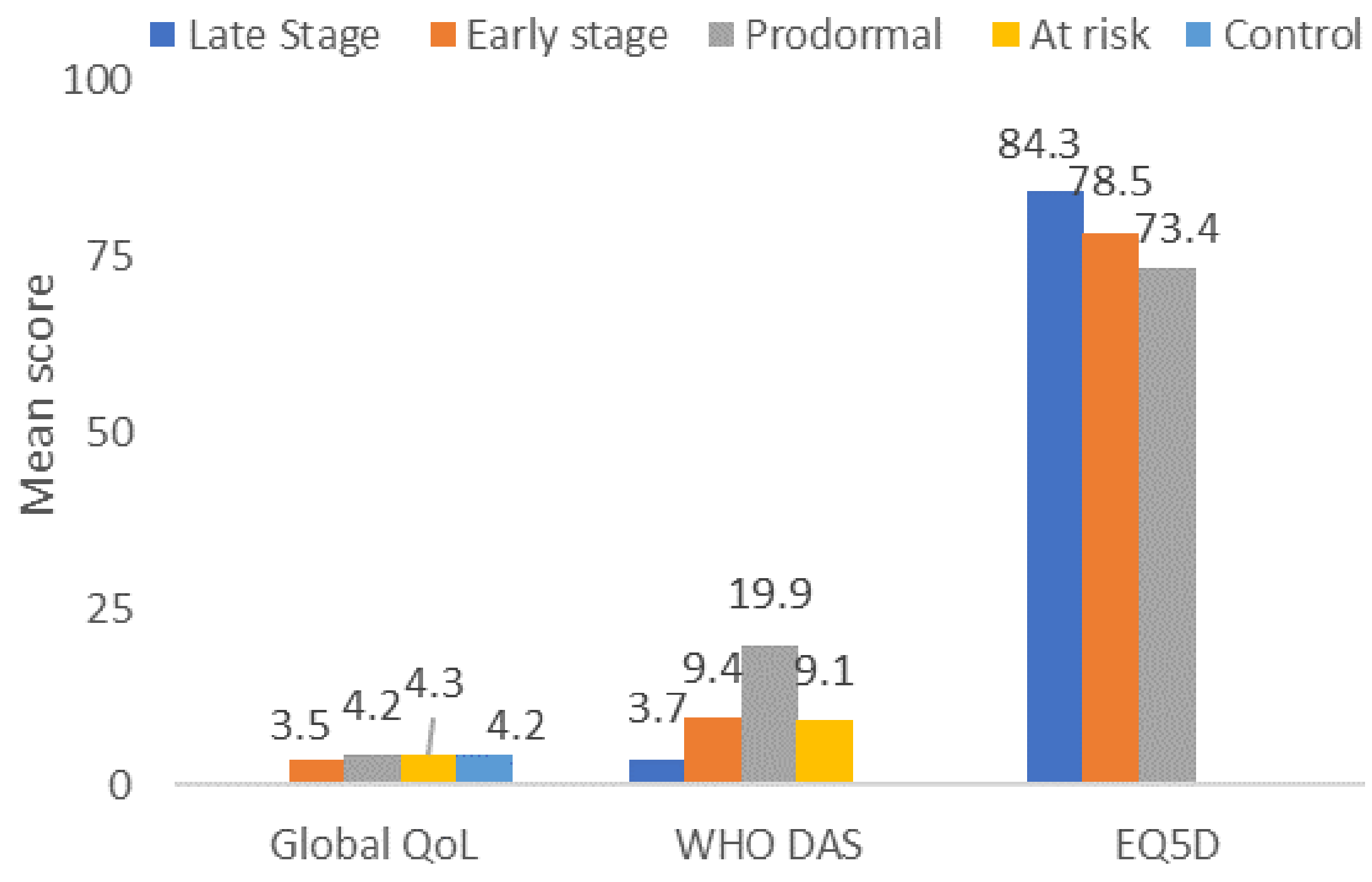
Records excluded
(n=92)
No SGA Disease (n=3)
Study design (n=68)
Country not of interest (n=21)

Included studies
(n=24 studies from 39 publications)

Results

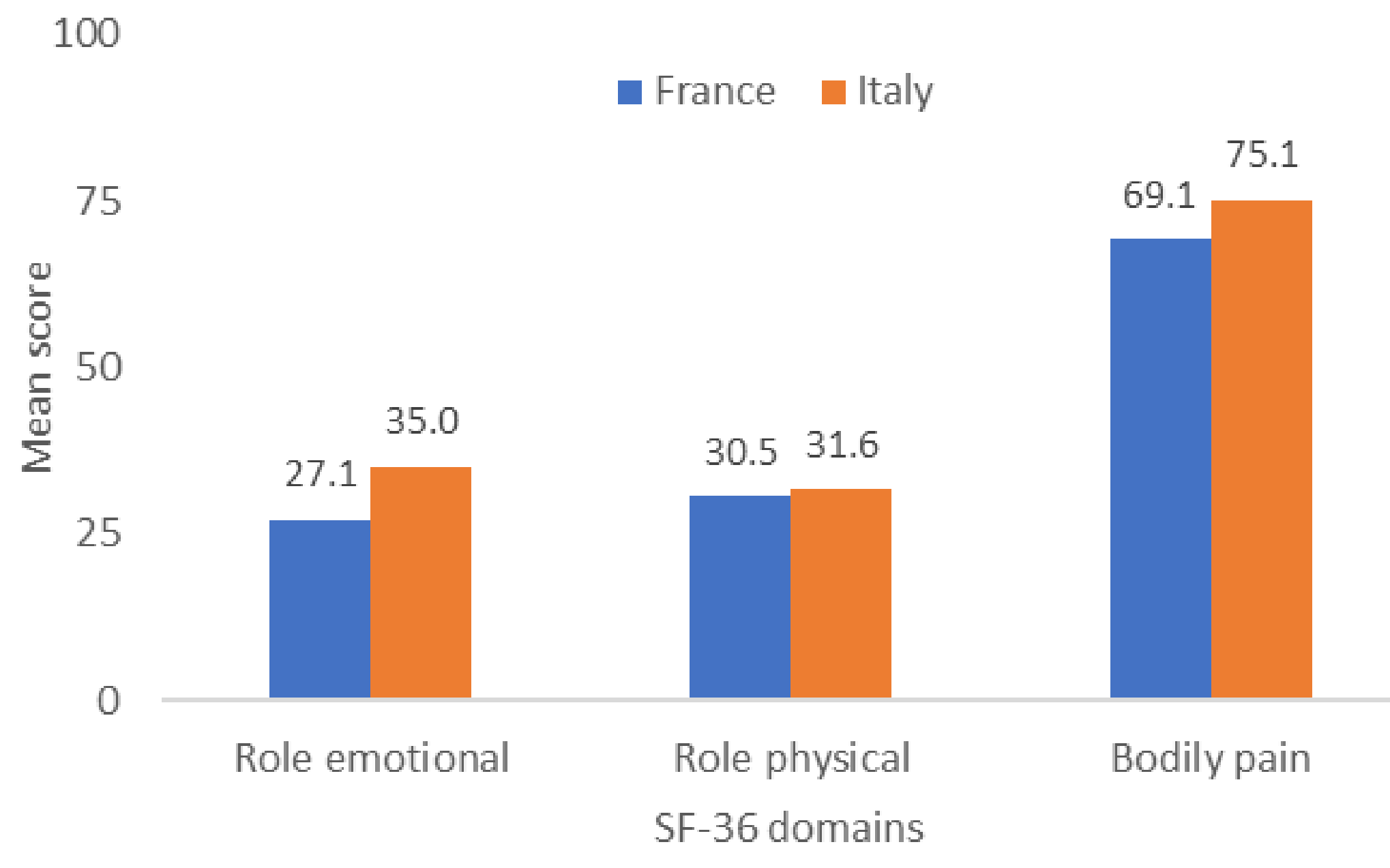
- Figure 1 summarizes the review process adopted to retrieve the 24 studies (EU4:7, US:10, UK:4, US/UK:2) assessing the humanistic burden of HD patients and caregivers
- HD patients (early and late stage) reported poor quality of life (Qol) on Global Qol, WHO-DAS, and EQ-5D scales compared to prodromal, at-risk, and control groups (Figure 2)[2,3,4]

Figure 2: Quality of life based on disease stage



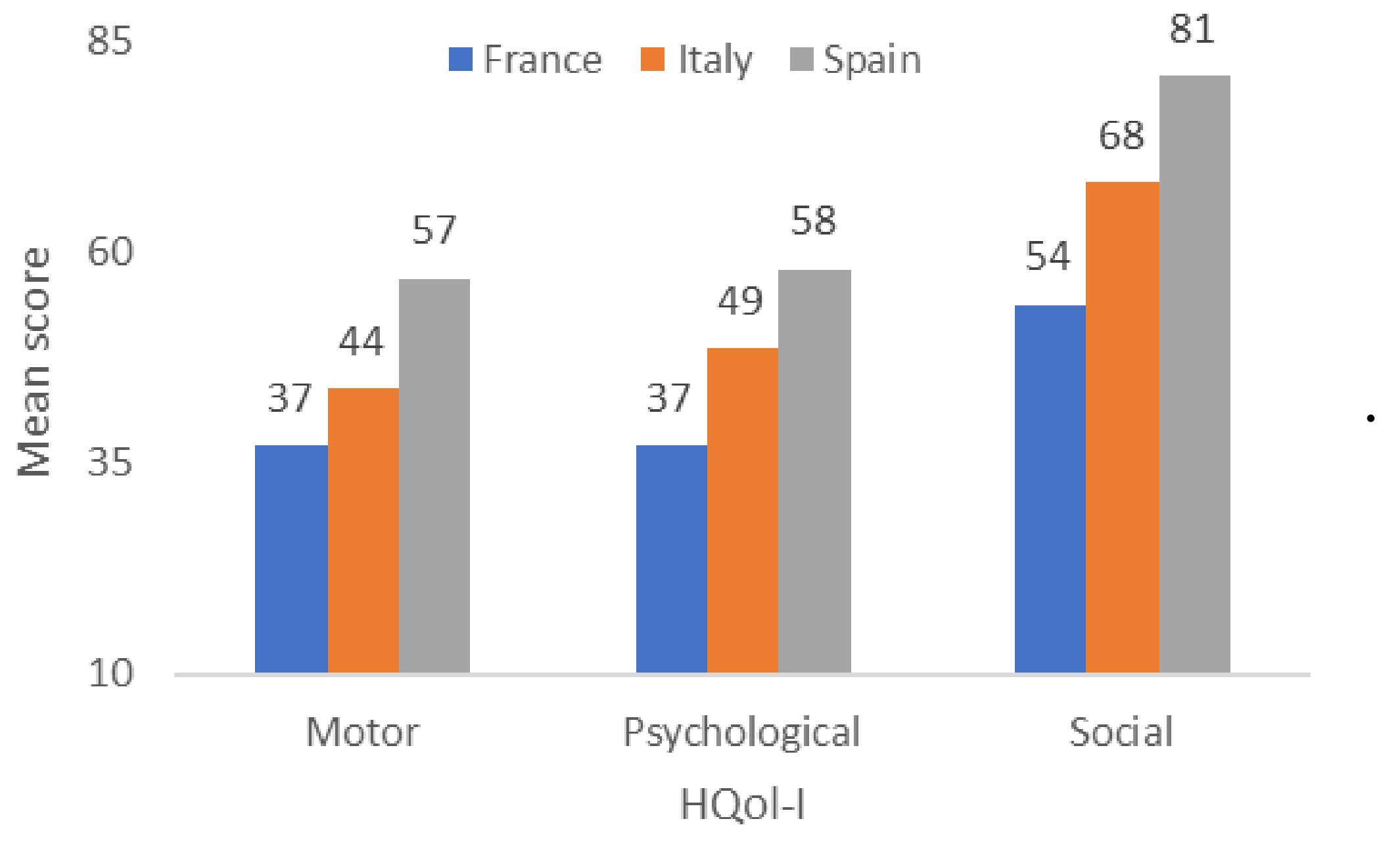
- The average mean (SD) chorea domain score of the HDQLIFE was 59.3 (6.1). Patients with later stage HD had, on average, higher Chorea domain scores (61.3) than those with early-stage HD (54.2)[5]
- Role emotional and role physical were the most altered dimensions on SF-36 (Figure 3)

Figure 3: Most altered domains on SF-36



- Motor (37.12 to 56.96) and psychological (37.02 to 58.06) were the most affected dimensions on the HQol-I scale (Figure 4), whereas self-care and usual activities were most altered on the EQ-5D scale [6,7,4]

Figure 4: Most altered domains on HQol-I



Results

- Bodily pain was the least altered dimension both on SF-36 (69.12 to 75.07) and EQ-5D scales, while the social dimension was the least altered on the HQol-I scale[6,7,8]
- Caregiver burden increased with the increase in severity of disease across Europe and US
- HD patients' functional scores and cognitive scores were found to be significant drivers of the caregiver burden
- Overall impairment in caregiver daily activities increased with disease severity: 42% in early stage, 48% in moderate stage and 52% in advanced stage [9]
- UK caregivers felt significantly more sad than US caregivers (group mean 7.14 vs. 5.39) while US caregivers worried significantly more about their finances (5.69 vs. 3.74; p=0.006), development of HD (3.95 vs. 0.00; p=0.003) and concerns about their children (7.05 vs 3.13 ; p=0.000) as compared to UK[10]
- The self-reported EQ-5D (58.38 ± 23.20 vs. 75.31 ± 20.31) and SF-12v2 (PCS: 41.52 ± 12.78 vs. 50.67 ± 8.94, MCS: 39.95 ± 11.50 vs. 43.85 ± 11.25) measures show that, as expected, patients scored worse than proxies on all indices of HRQoL[11]

Conclusions

Chorea negatively affected HRQoL. Substantially higher burden of HD has been observed on society, patients, and caregivers across Europe and US. Motor and psychological are the most affected domains on various Qol scales. Better treatment options are needed to improve motor function and thereby HRQoL of HD patients.

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Disclosure:

GK, PR, SK, BS, the authors, declare that they have no conflict of interest

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