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Letter to the Editor

# Pain in adult patients with Pompe disease

*To the Editor,*

We recently published an article in this journal entitled "Pain in adult patients with Pompe disease: A cross-sectional survey" (Güngör et al., Mol Genet Metab. 2013; (109):371–376) and would like to present some additional data from a similar study we were involved in.

A group of 25 patients from the United Kingdom (UK) were surveyed with the long form of the Brief Pain Inventory (BPI). This questionnaire asks patients to score pain within the previous 7 days, instead of the last 24 h assessed in the more commonly used short-form BPI applied in our earlier paper.

Eighty-eight percent of UK patients reported pain in the last week, a considerably higher proportion than the 45% reporting current pain in our earlier paper (Table 1). The pain severity scores (PSS) (3.9 (0.5–7.5) vs 3.1 (0.75–8.0) in Güngör et al.) and the median pain interference scores (PIS) (3.9 (0–7.5) vs 3.3 (0–8.4) in Güngör et al.) were only slightly higher.

The UK group did not differ in their demographic data from the patients in our earlier paper. The sites (shoulder girdle 76%, hip girdle 52%) and type of pain (dull/pressing 76%, exhausting 68%, unbearable 60%, tender 52%) were consistent with Güngör et al.. Pain mostly affected the limb girdle area, which is known to be one of the areas most affected by muscle weakness in this disease. Fifteen patients (60%) received treatment for pain.

The short-form BPI scores pain within the last 24 h to minimize recall bias, which has been shown to inflate the PSS and PIS scores. The present data suggest that, in addition to a large proportion of patients having current pain, an even larger group suffers from pain in daily life. These findings again emphasize the importance of pain in the long-term treatment of patients with Pompe disease.

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**Table 1**
Demographic and pain characteristics in 25 UK adult patients with Pompe disease. Data are given either as median (range) or numbers (percentage).

| Characteristic | Pompe patients (n = 25) |
|---|---|
| Median age, years (range) | 47 (20–70) |
| Female, n (%) | 13 (52) |
| Median age at first symptoms, years (range) | 25 (2–54) |
| ERT, n (%) | |
| Currently receiving | 19 (76) |
| Discontinuation | 2 (8) |
| Never | 4 (16) |
| Patients reporting pain in the last 7 days, n (%) | 22 (88) |
| *Pain severity score (PSS) (0–10)*[a] | |
| Median PSS (range) | 3.9 (0.5–7.5) |
| Pain severity subgroups | |
| No pain (rating of 0), n (%) | – |
| Mild pain (1–3), n (%) | 10 (45.5) |
| Moderate pain (4–6), n (%) | 10 (45.5) |
| Severe pain (7–10), n (%) | 2 (9) |
| *Pain related interference with daily activities* | |
| Median pain interference score (range) | 3.9 (0–7.5) |
| General activity, median (range) | 4.0 (0–10) |
| Mood, median, (range) | 3.0 (0–10) |
| Walking ability, median (range) | 5.0 (0–10) |
| Normal work, median (range) | 4.0 (0–10) |
| Relations with other people, median (range) | 3.0 (0–10) |
| Sleep, median (range) | 4.0 (0–10) |
| Enjoyment of life, median (range) | 4.0 (0–10) |
| *Quality of life/other patient reported outcomes* | |
| Median HADS depression score (range) | 8.0 (0–14) |
| Median HADS anxiety score (range) | 7.0 (1–18) |
| Median SF-36v2 physical component summary score (range) | 27 (14–55) |
| Median SF-36v2 mental component summary score (range) | 48 (24–71) |
| Median Rotterdam Handicap Scale Score (range) | 24 (12–36) |

HADS — Hospital Anxiety and Depression Score, SF-36v2 — Short Form Health Survey 36 version 2.
[a] Pain was scored within the last 7 days.

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