CLINICAL ASSESSMENT

The Pain in Dystonia Scale (PIDS)—Development and Validation in Cervical Dystonia

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ABSTRACT: Background: A better understanding of pain in adult-onset idiopathic dystonia (AOID) is needed to implement effective therapeutic strategies.
Objective: To develop a new rating instrument for pain in AOID and validate it in cervical dystonia (CD).
Methods: Development and validation of the Pain in Dystonia Scale (PIDS) comprised three phases. In phase 1, international experts and participants with AOID generated and evaluated the preliminary items for content validity. In phase 2, the PIDS was drafted and revised by the experts, followed by cognitive interviews to ensure self-administration suitability. In phase 3, the PIDS psychometric properties were assessed in 85 participants with CD and retested in 40 participants.
Results: The final version of PIDS evaluates pain severity (by body-part), functional impact, and external modulating factors. Test–retest reliability showed a high-correlation coefficient for the total score (0.9, $P < 0.001$), and intraclass correlation coefficients were 0.7 or higher for all items in all body-parts subscores. The overall PIDS severity score showed high internal consistency (Cronbach's α, 0.9). Convergent validity analysis revealed a strong correlation between the PIDS severity score and the Toronto Western Spasmodic Torticollis Rating Scale

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pain subscale (0.8, $P < 0.001$) and the Brief Pain Inventory-short form items related to pain at time of the assessment (0.7, $P < 0.001$) and impact of pain on daily functioning (0.7, $P < 0.001$).

Conclusion: The PIDS is the first specific questionnaire developed to evaluate pain in all patients with AOID, here, demonstrating high-level psychometric properties in people with CD. Future work will validate PIDS in other forms of AOID. © 2023 International Parkinson and Movement Disorder Society.

Key Words: cervical dystonia; pain; scale development; scale validation; measurement tool

Introduction

Adult-onset isolated dystonia (AOID) is associated with a complex spectrum of non-motor symptoms, in particular depression, anxiety, and pain, which influence considerably patients' health-related quality of life and daily functioning.[1-4] Although non-motor features vary across the different focal forms of AOID (cervical, cranial, laryngeal, and upper limb), pain is reported in all these forms, manifesting at highest frequency in cervical dystonia (CD),[5-9] which is the most common, and characterized by abnormal posturing and movements of head, neck, and shoulders. Pain is its most frequent non-motor symptom, with a prevalence ranging from 54.6% to 88.9%.[10] Mainly perceived in the neck and shoulders, it often spreads to the upper back region and sometimes radiates cranially on the side of head deviation and caudally to the ipsilateral upper limb.[11] Approximately 10% to 20% of CD patients suffer from different clinical forms of headache.[7] Among the other AOID, people with blepharospasm complain of ocular dysesthesia and photo-oculodynia, and more than a third of patients with upper or lower limb dystonia report clinically significant pain.[12] The mechanisms of pain in AOID remain incompletely understood and might include both musculoskeletal and central pain-modulating systems.[7,13-15]

Better recognition and understanding of pain in AOID are needed to implement effective therapeutic strategies. For this, validated and specific screening and severity rating scales are needed. To date, there is uncertainty on how to assess pain in AOID, and a surprising dearth of validated rating instruments.[16] To the best of our knowledge, the only instruments developed to measure pain in this disorder are the pain subscales of the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) and of its newer version included in the Comprehensive Cervical Dystonia Rating Scale,[17] which are applicable only to CD. This subscale considers exclusively neck pain and is comprised of subscores for severity, duration, and related disability, all using a 0–5 Likert rating. By assessing pain only in the body region affected by dystonic muscle contractions, this instrument is skewed toward a musculoskeletal conceptualization of pain and cannot evaluate a more general dysfunction of nociceptive systems in AOID. Moreover, a relevant proportion of patients with CD have dystonia in other body regions that can also present with associated pain.

Existing phenomenological descriptions, and the potential involvement of different pain-generating mechanisms related to the core sensorimotor processing alterations of dystonia, support the rationale for developing a novel disease-specific instrument to assess pain in AOID. Our approach to pain measurement in AOID aims to overcome the existing limitations through the development of a self-administered instrument (hence, of more practical applicability) that has the following characteristics: (1) self-rating of pain intensity using a visual analog scale instead of a Likert-type rating; (2) evaluation of pain across different body regions; (3) a more granular assessment of the impact of pain on daily living activities; (4) complementing pain severity rating with the assessment of external factors that can trigger or alleviate pain.

Here, we present the development of this new instrument and its initial validation. Given the relevant clinical and pathophysiological differences across different forms of AOID, we started the validation of the scale in the most common form of AOID, CD.

Methods

This was a cross-sectional, single-center, one-point-in-time evaluation with retest comprised of three phases: item development, scale development, and scale evaluation.

Patients and Consent

Patients with CD diagnosed according to international consensus criteria attending the Movement Disorders Clinic, University of Calgary, were invited to participate. Exclusion criteria included: inability to give consent, dementia formally diagnosed according to Diagnostic and Statistical Manual of Mental Disorders-5 criteria[18] or diagnosis of disorders causing pain clearly unrelated to dystonia (eg, severe osteoarthritis/arthritis, malignancy). All participants provided informed consent. This study was approved by the University of Calgary Research Ethics Board (REB19-2111).

Phase 1: Scale–Item Development

Domain Delineation and Item Generation

The purpose of this first phase was to specify pain in dystonia as the primary scale domain, to define its boundaries, and to identify appropriate questions fitting the chosen domain. We aimed to assess not only localization, intensity, and frequency of pain, but also the relationship between pain and functional impact on daily living activities, as well as external factors aggravating or relieving pain severity. This was done by first confirming that no existing instruments addressed the scale subject. Next, the lead authors (V.B. and D.M.) generated an initial list of items and questions suitable to explore the chosen domain.

Development of Item Pool

Feedback on how the proposed items to address the domain of interest were sought from a group of patient representatives and an international panel of experts (F.M., R.E., S.F., A.S., M.S., M.J.E., S.C., G.D., K.R.C., S.P.R., and H.J.). First, the expert panel evaluated each item for content relevance, representativeness, and practical quality. A questionnaire was then administered to 10 people with CD, enrolled consecutively from the Calgary Movement Disorders outpatient clinic, and to the international panel of experts who rated each item on whether the domain measured was essential/valuable in assessing pain in CD. Both people with CD and experts were also asked to suggest other items that the researchers had not yet proposed. The importance of each item was weighted by the content validity ratio (CVR) to assess the rank of each item as judged by the panel of raters. Patients were asked to rate each item as essential, valuable, or not valuable to assess pain in the general population of people with CD. Each clinical expert's panel member expressed the same rating considered the following: (1) "Does this clinical feature help substantially in measuring pain?"; (2) "Is assessing this clinical feature sufficiently reliable to be applied on a large clinical scale?"; (3) "Is the assessment of this clinical feature practically feasible as well as sufficiently time- and cost-effective to be applied on a large clinical scale?". The CVR was measured using the following formula: $CVR = (1\text{-}NE/2)/(N/2)$, where NE = number of raters indicating "essential" and N = total number of raters. Any item that fell below a CVR of 0.6 was removed. Items were retained only if they had passed the content validity assessment in both the expert and patient representatives' panel. This procedure provided the final list of the rating scale items.

Phase 2: Scale Development

After the final selection of items, V.B. and D.M. developed the first version of the scale with a proposed grading system and a weighting system for scoring based on the individual items' CVR values and their clinical experience. A Likert-type of grading was favored when possible, avoiding exceeding four anchor points per item (including "0", always indicating the absence of the clinical feature) to enhance the scale's consistency. The scale structure underwent three iterative revisions by the international expert panel until an agreement was reached.

After this step was completed, V.B., B.A., and D.M. operationalized the questions' pre-testing and the scale's survey administration procedure. Each item was assessed through fixed questions the patient read during a self-administration procedure using cognitive interviewing. Thirteen respondents were asked to verbalize the mental process of providing their answers. This approach helped to determine whether the questions were generating the information intended to be obtained by helping to ensure that respondents understood the questions as initially envisioned. Item reduction was performed, based on the number of correctly understood questions per item and the probability of a particular examinee understanding correctly a given item. The results of this phase led to the final version of the self-administered scale (named Pain in Dystonia Scale [PIDS]) that was used for evaluation.

Phase 3: Scale Evaluation

Participants in this phase were assessed clinically, obtaining their demographic variables (age, sex, and comorbid medical conditions) and disease-related variables (duration of dystonia, family history, and current dystonia treatment). They completed a self-administered final version of the PIDS (the first 40 participants were requested to complete the final version of the scale for a second time to assess stability).

In addition, the following assessments were applied.

Cervical Dystonia Validated Assessment Tools

- TWSTRS[19] pain subscale, consisting of three patient-rated items, two of which are scored on a range from 0 to 5, while the third depends on patients' score of their usual pain (factored by 2), worst pain, and best pain, on a range of 0 to 10, all divided by four to reach a total ranging from 0 to 10 (maximum score of 20). We selected the pain subscale of the earlier TWSTRS, rather than that of the TWSTRS-2,[20] because of its longer history of use.
- TWSTRS psychiatric screening tool (TWSTRS-PSYCH),[21] including six items assessing depression, loss of interest, discomfort, anxiety, physical symptoms of panic attack, and afraid of going outside. The total score is a sum of all items (maximum score 24).

- Global Dystonia Severity Rating Scale (GDRS),[22] serving as an instrument to assess dystonia severity. The total score is the sum of the scores for all the body regions (maximum total score 140).
- Cervical Dystonia Questionnaire 24 (CDQ-24),[9] evaluating quality of life in patients with CD and blepharospasm. It includes 24 items based on five subscales (stigma, emotional wellbeing, pain, activities of daily living, and social/family life). Each item consists of five statements representing increasing severity of impairment, scored from 0 to 4 (maximum score 96).

Pain Assessment Tools

- Brief Pain Inventory Short Form (BPI-sf),[23] a 9-item self-administered questionnaire used to evaluate the severity of a patient's pain and the impact of this pain on the patient's daily functioning. Answers are rated from 0 (no pain) to 10 (pain as bad as you can imagine). The impact on daily functioning answers range between 0 (does not interfere) and 10 (completely interferes). Items are rated individually.
- American chronic pain association quality-of-life scale (QOLS)[24] for people with pain, a brief measure of function for people with pain (from 0-: "Stay in bed all day/feel hopeless about life" to 10 "Go to work/volunteer each day. Normal daily activities each day. Have asocial life outside of work. Take an active part in family life").

Sample Size and Statistical Analysis

The sample size was calculated using an 8 to 10:1 sample-to-item ratio shown to be adequate for this analysis.[13] As we aimed to develop a user-friendly tool with <10 items, a sample size of 80 subjects was considered appropriate. With respect to evaluation of stability, several methodological guidelines[25-27] suggest that a minimum sample size of 30 individuals is considered appropriate for a test–retest analysis. To guarantee sufficient accuracy to our analysis, we opted to include the first 40 participants in this analysis.

Data was collected and entered in an anonymized database. Statistical analysis was performed using STATA v. 16. In addition to descriptive statistics, the following psychometric features were assessed:

1. Test–retest reliability: calculated with intraclass correlations (ICCs) based on 2-way mixed-effects models, average measures, and absolute agreement. An ICC ≥0.7 was considered satisfactory.
2. Acceptability: calculated using missing responses, with <5% considered acceptable.
3. Distribution: based on observed means versus median scores. A difference of <10% of the maximum possible scale score was considered acceptable.
4. Floor and ceiling effects: taking 15% as the maximum and minimum acceptable.
5. Internal consistency: a Cronbach's α item was used to assess internal validity considering 0.7 or greater acceptable.
6. Validity: correlation coefficients between the PIDS score and existing CD or pain assessment tools were calculated with Spearman's ρ. Based on its strength, the correlation was classified as weak ($r \leq 0.4$), moderate ($0.4 \geq r \leq 0.7$), or strong ($r \geq 0.7$).

Results

Ten participating people with CD contributed to the development of the item pool in phase 1, 13 participated in the cognitive interviews in phase 2, and 85 completed the final version of the scale and additional assessments in phase 3 (the first 40 also participated in the test–retest assessment). Descriptive data are shown in Table 1.

The study was conducted during the coronavirus disease 2019 (COVID-19) pandemic. Interviews were performed virtually using teleconference software. Participants completed an electronic version of the scale using the UCalgary Survey tool powered by QualtricsXM. The median time for completion was 10.4 minutes (range, 3.3–20.9 minutes).

The final version of the scale included three sections severity of pain, functional impact, and external modulating factors (Fig. 1). The instrument assesses the frequency and severity of pain experienced by individuals within the previous week.

Section 1: Pain Severity

Participants could complete the scale per body part affected by pain including neck and shoulders, eyes, jaw, arms, legs, and mid/lower back.

This section included three items: (1) pain intensity at its worst (0–10); (2) pain intensity on average (0–10); and (3) days × week with pain (adopting a rating from 0–3: 0 = no pain; 1 = <1 day/week; 2 = 2–4 days/week; 3 = >5 days/week). A free text section was included for patients to report an average amount of hours with pain in the days in which they experienced it. Section 1 subscores per body part were computed as follows: (pain intensity on average *2 + pain intensity at its worst)/3 * days/week score 0–3. This generates a total subscore ranging from 0 to 30. Section 1 subscores per body part were added to obtain the final score. The psychometric validation of the scale was performed on this section.

Section 2: Functional Impact

Participants were asked to rate the degree of impact that pain has on different activities including engaging in physical exercise, participating in social events and gatherings, completing household activities, driving,

TABLE 1 . *Demographics and clinical characteristics of the validation study population*

Demographics	
Female, No. (%)	66 (77.7)
Age in years–mean ± SD	61.7 ± 10.1–range, 31–82
Disease duration in years–mean ± SD	12.5 ± 10.5–range, 1–41
Age at onset in years–mean ± SD	49.2 ± 12.7–range, 16–77.5
Married, No. (%)	62 (74.7)
Receiving botulinum toxin treatment (%)	78 (91.8)
Self-reported family history of dystonia (%)	13 (15.3)
Post-secondary education (%)	70 (82.7)
Descriptive statistics of the assessments in the study	
TWSTRS pain subscale–mean ± SD	16 ± 10.5
TWSTRS psychiatric screening tool–mean ± SD	5.8 ± 4.9
GDS–mean ± SD	4.8 ± 3.5
CDQ24–mean ± SD	26.9 ± 17.8
BPI-sf, pain at its worst in the last 24 hours (item 3)–mean ± SD	4.6 ± 3
BPI-sf, pain at its least in the last 24 hours (item 4)–mean ± SD	1.9 ± 2.4
BPI-sf, pain on the average (item 5)–mean ± SD	3.7 ± 2.5
BPI-sf, pain right now (item 6)–mean ± SD	3.1 ± 2.7
BPI-sf, impact of pain on daily functioning (item 9 a–f)–mean ± SD	18.8 ± 17.4
American chronic pain association QUOLS score–mean ± SD	7.8 ± 2.1
Reported pain, No. (%)	
No pain (%)	12 (14.1)
Pain in neck and shoulders (%)	72 (84.7)
Pain in eyes	29 (34.1)
Pain in jaw	29 (34.1)
Pain in arms	29 (34.1)
Pain in legs	21 (24.7)
Pain in mid/lower back	39 (45.9)
Pain in 1 body part	12 (14.1)
Pain in 2 body parts	19 (22.4)
Pain in 3 body parts	16 (18.8)
Pain in 4 body parts	15 (17.7)
Pain in 5 body parts	5 (5.9)
Pain in 6 body parts	6 (7.1)

Abbreviations: SD, standard deviation; TWSTRS, Toronto Western Spasmodic Torticollis Rating Scale; GDS, Global Dystonia Severity Rating Scale; CDQ-24, Cervical Dystonia Questionnaire 24; BPI-sf, Brief Pain Inventory short form; QOLS, Quality of Life Scale.

getting a good night sleep or rest, outdoor leisure activities, working, and personal relationships. Responses are provided on a Likert scale from 0 to 3: 0 = no interference, 1 = sometimes interferes, 2 = often interferes, and 3 = unable to perform this because of pain.

Section 3: External Modulating Factors

Participants were asked about external factors that could have an aggravating or relieving effect on their pain. The section is divided in the two following items:

2.a. External factors that can trigger pain included heat, cold or both, bright lights or changes in lighting, exercise, manipulation or massage, changes in posture (eg, standing, sitting, or lying down), time of day, stress, and prolonged fixed position. Responses are provided on a Likert scale from 0 to 3: 0 = no effect, 1 = mild effect, 2 = moderate effect, and 3 = severe effect.

1. SEVERITY

Which body parts are most affected by pain? How often do you experience pain in each body part?
We are interested in your experience over the **past week** including today.

How often do you experience pain in your **neck and shoulders?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to neck and shoulder pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to neck and shoulder pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your neck and shoulders, how many **hours** do you suffer pain on average? _____
How often do you experience pain or other uncomfortable sensations around your **eyes?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to eye pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to eye pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your eyes, how many **hours** do you suffer pain on average? _____
How often do you experience pain in your **jaw?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to jaw pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to jaw pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your jaw, how many **hours** do you suffer pain on average? _____
How often do you experience pain in your **arms?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to arm pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to arm pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your arms, how many **hours** do you suffer pain on average? _____
How often do you experience pain in your **legs?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to leg pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to leg pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your legs, how many **hours** do you suffer pain on average? _____
How often do you experience pain in your **mid lower back?** ☐ No pain in this body part ☐ Less than 1 day per week ☐ 2-4 days per week ☐ 5 or more days per week	Please indicate pain at **its worst** in regards to mid lower back pain? No Pain _____ Worst Pain Imaginable Please indicate pain **on average** in regards to mid lower back pain? No Pain _____ Worst Pain Imaginable On the days that you experience pain in your mid lower back, how many **hours** do you suffer pain on average?_____

FIG. 1. Pain in dystonia (PIDS)–assessment tool.

2. FUNCTIONAL IMPACT

Pain can impact daily life activities. Please rate the degree that **PAIN** has on the following activities.

	N/A	No interference (0)	Sometimes interferes (1)	Often interferes (2)	Unable to perform this due to pain (3)
Engaging in physical exercise					
Participating in social events and gatherings					
Completing household activities i.e., cooking, leaning.					
Driving					
Getting a good night sleep or rest					
Outdoor leisure activities					
Working					
Personal relationships					

3. EXTERNAL FACTORS

Some external factors can **trigger pain** or make it worse. Using the scale, please indicate the degree to which these factors affect you.

	N/A	No effect (0)	Mild effect (1)	Moderate effect (2)	Severe effect (3)
Heat or cold or both					
Bright lights or changes in lighting					
Exercise					
Manipulation or Massage					
Changes in posture (e.g., standing, sitting or lying down)					
Time of day					
Stress					
Prolonged fixed position					

FIG. 1. (Continued)

Some external factors can provide **relief of pain**. Using the scale, please indicate the degree to which these strategies improve your pain.

	N/A	No relief (0)	Mild relief (1)	Moderate relief (2)	Complete relief (3)
Heat or cold or both					
Physical rest					
Exercise					
Sleep					
Manipulation or Massage					
Stretching					
Relaxation techniques					
Actions/gestures you do to alleviate dystonia *(these are actions used by some patients to alleviate dystonia i.e resting your head on the headrest, touching chin or face, massaging your eyes or other actions)*					
Changes in posture (e.g. standing, sitting or lying down)					
Alcohol					
Self prescribed treatments					

FIG. 1. (Continued)

2.b. External factors that can relieve pain included heat, cold or both, physical rest, exercise, sleep, manipulation or massage, stretching, relaxation techniques, actions/gestures done to alleviate dystonia, changes in posture, alcohol, and self-prescribed treatments. Responses are provided on a Likert scale from 0 to 3: 0 = no relief, 1 = mild relief, 2 = moderate relief, and 3 = complete relief.

Reliability, Acceptability, and Distribution

Test–retest reliability was examined in a consecutive series of 40 patients assessed on two occasions, with a median of 9 days (range, 7–17) days' separation. The re-test was completed within 7 days of the treatment with botulinum toxin, if applicable, to avoid the confounding effect of the treatment on the results. The test–retest reliability showed a significant correlation coefficient for the total score (0.9, $P < 0.001$), and intraclass correlation coefficients were 0.7 or higher for all items in all sub scores by body part (Table 2).

There were no missing data as the scale was designed and implemented to avoid the chance of leaving incomplete answers or blanks. Twelve participants did not report pain in any body part. The median, interquartile, and full range for the PIDS score were 21.6, 8.1–39.5, and 0.4–125.5. The distribution was mildly skewed (1.5). The difference between observed mean and median scores was between 10% of the maximum possible scale score for all subscores by body part, demonstrating acceptable distribution. Median scores and full ranges are shown in Figure 2. Differences in the distribution of the final score were based on the number of body parts affected by pain. Among those patients with pain, there was no floor (9.6% responses below the 10th percentile) or ceiling effect (8.2% responses above the 90th percentile).

TABLE 2 *PIDS test–re-test intraclass correlations coefficients and 95% confidence intervals for severity score items by body part*

	Neck and shoulders	Eyes	Jaw	Arms	Legs	Mid/lower back
Pain on average	0.8 (0.7–0.9)	0.8 (0.6–0.9)	0.9 (0.7–1)	0.7 (0.4–0.9)	0.9 (0.8–1)	0.8 (0.7–0.9)
Pain at its worst	0.9 (0.8–0.9)	0.7 (0.5–0.9)	0.8 (0.6–0.9)	0.7 (0.4–0.9)	0.9 (0.8–1)	0.7 (0.5–0.9)
Days × week	0.8 (0.6–0.9)	0.9 (0.8–0.9)	0.9 (0.8–1)	0.7 (0.6–0.9)	0.7 (0.6–0.9)	0.9 (0.9–1)

Abbreviation: PIDS, pain in dystonia scale.

Internal Consistency

The overall PIDS had a high internal consistency measured by Cronbach's α (0.9). The internal consistencies for the subscores by body part affected by pain were excellent (Cronbach's α, 0.9 for neck and shoulder pain, 0.9 for eye pain, 1.0 for jaw pain, 1.0 for pain in the arms, 1.0 for pain in the legs, and 0.9 for middle back pain).

The functional impact and external modulating factors sections also showed high internal consistency (Cronbach's α, 0.9 for both items independently).

Validity

The assessment of convergent validity revealed a strong correlation between the PIDS and the TWSTRS pain subscale (0.8, $P < 0.001$), the BPI-sf items related to pain at the moment of the assessment (0.7, $P < 0.001$), and impact of pain on daily functioning (0.7, $P < 0.001$), and moderate correlations with the BPI-sf items related to pain at its worst in the last 24 hours (0.6, $P < 0.001$) and to pain on average (0.7, $P < 0.001$), as well as with the CDQ-24 (0.5, $P < 0.001$). In addition, there was a mild significant positive correlation between the PIDS and GDRS (0.4 $P = 0.007$) and TWSTRS-PSYCH (0.3, $P = 0.001$) scores, as well as a significant negative correlation between the PIDS and the American chronic pain association QOLS (−0.5, $P < 0.001$) (Table 3).

Discussion

We report the development of the first self-administered scale for a comprehensive evaluation of pain across all body regions in AOID, and here, validated in CD. Scale items were developed with the contribution of people living with CD. The PIDS is easy to administer also remotely, with a median completion duration of 10 minutes. We demonstrated excellent test–retest reliability, high internal consistency, and strong convergent validity, measures in respect to disease-specific and general pain rating instruments and instruments rating dystonia severity and health-related quality of life.

Different body locations of dystonia may differ in non-motor symptoms and in their underlying pathophysiology.[13] We addressed the validation of this new scale adopting a "splitting" approach, first conducting a validation study in CD. Our sample is representative of the general population of patients with CD attending a standard movement disorders outpatient clinic

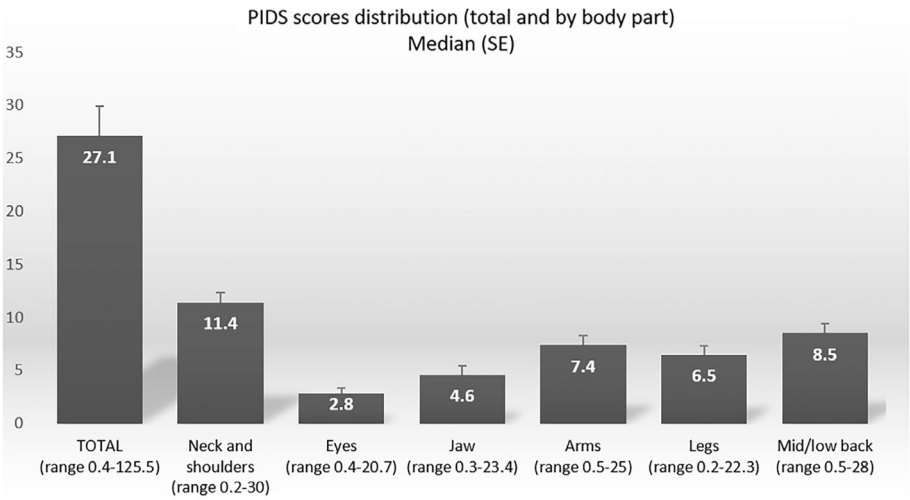

FIG. 2. Pain in dystonia (PIDS) scores distribution (total and by body part).

TABLE 3 *PIDS validity assessment*

	Correlations (Spearman ρ; <0.001)*
TWSTRS pain subscale	0.8*
TWSTRS psychiatric screening tool	0.3*
GDS	0.4*
CDQ24	0.5*
VAS	0.6*
BPI-sf, pain at its worst in the last 24 hours (item 3)	0.6*
BPI-sf, pain on the average (item 5)	0.7*
BPI-sf, pain right now (item 6)	0.7*
BPI-sf, impact of pain on daily functioning (item 9 a–f)	0.7*
American chronic pain association's QOLS	−0.5*

Abbreviations: PIDS, pain in dystonia scale; TWSTRS, Toronto Western Spasmodic Torticollis Rating Scale; GDS, Global Dystonia Severity Rating Scale; CDQ-24, Cervical Dystonia Questionnaire 24; VAS, visual analogue scale; BPI-sf, Brief Pain Inventory short form; QOLS, Quality of life scale.
*$P < 0.001$.

suggesting possible generalizability of application in this form of dystonia. Patients were tested after adequate washout from botulinum toxin, which is the most common treatment for CD, although residual or persistent anti-nociceptive effects of this cannot be completely excluded.

Unlike the TWSTRS pain subscale, which focuses on the neck, the PIDS provides a comprehensive self-evaluation of pain from six different body regions, likely encompassing different types of pain in AOID. Its total score provides the overall sum of the burden of pain in dystonia, similar to other disease-specific pain scales (eg, the King's Parkinson's disease pain scale).[28]

In developing the new scale, we avoided a Likert rating for pain intensity because of its limited number of values and its dependence on subjective interpretation of its anchor points. Despite some evidence suggesting that a numerical rating scale (0–10) is equally valid, but easier to use than a visual analog scale (VAS) to assess pain,[29,30] we chose a VAS-based assessment as it better reflects general aspects of pain intensity with its character and affective perception, more in line with the conceptualization of pain as a complex biopsychosocial experience affecting quality of life.[31] We developed a self-administered tool to eliminate rater bias and used clear instructions, neutral response options, and monitored missing data to reduce social desirability or response bias. We selected validated and standardized scales and used multiple measures to the same construct to mitigate systematic measurement errors. The functional impact subscale includes a broad series of daily routine activities, supporting its applicability to different forms of AOID. Although not included in a scoring process, collecting information on the influence of external factors on pain experience could improve characterization and clinical interpretation of pain, maximizing accuracy during follow-up and when interpreting treatment response.

Our analysis of scale attenuation effects for the pain severity subscale was reassuring, demonstrating absence of any relevant ceiling or floor effect. Its internal consistency was also high for both overall score and for individual body region subscores. Internal consistency was also very high for the functional impact subscore and the section exploring external modulating factors.

The significant correlation of the PIDS score to severity of CD (TWSTRS severity subscore) and of dystonia across the whole body (GDRS) supports the existence of a relationship between pain and the severity of motor symptoms in dystonia. Likewise, the significant correlation of the PIDS score with the CDQ24 indicates an influence of pain on factors determining disease-specific health-related of quality of life in this patient population. Importantly, the scale did not show significant sex differences or a significant effect of education, which provides further support to the generalizability of its validity to the whole population of patients with CD. Test–retest reliability was excellent for all items and body regions with respect to the pain severity subscale. The time interval between the two time points of the test–retest assessment was kept as short as possible to minimize any potential influence of botulinum toxin treatment on either time point.

This new scale allows comparison of the frequency of occurrence and body distribution of pain symptoms between different forms of AOID and between these and control groups. This is relevant, given that different mechanisms may underlie pain in AOID. Although a musculoskeletal contribution is plausible, pain can occur in non-dystonic muscles, pressure algometry showed no stringent correlation with degree of contraction of dystonic muscles[32] and, in some patients, motor improvement with botulinum toxins and deep brain stimulation is not paralleled by pain improvement, suggesting a central contribution. Evidence suggests preserved pain threshold and tolerance and ascending nociceptive pathways, whereas a possibly primary deficit of the endogenous descending inhibitory pain system, relying on a spino-bulbo-spinal loop, has been documented in CD, but not in blepharospasm.[15] The data collected by this instrument could guide future research toward better pathophysiological subtyping of pain in AOID and of how this relates to motor and other non-motor features of dystonia. Future insights into the classification of pain in AOID might lead to adaptations to this instrument.

We acknowledge a few limitations of this study. The PIDS evaluates pain-related symptoms in AOID "lumping" different pain manifestations. This etiology-agnostic approach combines pain symptoms that are mechanistically related to dystonia and pain symptoms that are independent of dystonia. Despite this potential limitation this approach allows addressing the global burden of pain-related symptoms at the same time in the same patient and its region-specific burden. Body region-specific pain can, therefore, be further assessed and managed as needed, tailoring workup and treatment on everyone. The time elapsed between botulinum toxin treatment sessions and the second rating used to test reliability (median, 9 days) might have confounded test–retest stability. However, any change ensued because of confounding therapeutic effect would have diminished stability, which was nevertheless excellent. Finally, information on responsiveness to change is lacking; future research should evaluate responsiveness to general analgesics as well as dystonia-related treatments such as botulinum toxin and deep brain stimulation.

In conclusion, we present validation data for the PIDS in a population of participants with CD, product of an international collaboration with experts and people living with CD. The PIDS showed excellent psychometric characteristics and may become a valuable tool in clinical practice, providing for the first time a comprehensive evaluation of pain in AOID. Future steps will include validation in other types of AOID. ■

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Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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