A new short and simple health-related quality of life measurement for paediatric rheumatic diseases: initial validation in juvenile idiopathic arthritis

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Abstract

Objective. To develop and validate a new short and simple measure of health-related quality of life (HRQL) in children with juvenile idiopathic arthritis (JIA).

Methods. The Paediatric Rheumatology Quality of Life Scale (PRQL) is a 10-item questionnaire that explores HRQL in two domains: physical health (PhH) and psychosocial health (PsH). Validation of the parent proxy report and child self-report versions of the instrument was accomplished by evaluating 472 JIA patients and ~800 healthy children. Validation analyses included assessment of feasibility, face and content validity; construct and discriminative ability; internal structure and consistency; test–retest reliability; responsiveness to clinical change; and minimal clinically important difference.

Results. The PRQL was found to be feasible and to possess both face and content validity. The PRQL score correlated in the predicted range with most of the other JIA outcome measures, thereby demonstrating good construct validity, and discriminated well between different levels of disease severity. Assessment of internal structure (factor analysis) revealed that the PhH and PsH subscales identify two unambiguously separated domains. The internal consistency (Cronbach’s α) was 0.86. The intraclass correlation coefficient for test–retest reliability was 0.91. The PRQL revealed fair responsiveness, with a standardized response mean of 0.67 in improved patients. Overall, the PRQL appeared to be more able to capture physical HRQL than psychosocial HRQL.

Conclusion. The PRQL was found to possess good measurement properties and is, therefore, a valid instrument for the assessment of HRQL in children with JIA. This tool is primarily proposed for use in standard clinical care.

Key words: Rheumatic diseases, Health-related quality of life, Paediatrics, Children, Adolescents.

Introduction

Paediatric rheumatic diseases (PRDs) are chronic multisystem inflammatory conditions that may influence many aspects of a child’s life, including not only the physical but also the social, emotional, intellectual and economic [1]. Therefore, a complete assessment of children with PRD requires an understanding of the impact of the disease, its complications and its treatment on a child’s life. Assessment of health-related quality of life (HRQL) is increasingly recognized as a fundamental component of...
the clinical evaluation of children with PRD [2–5]. It has been suggested that measurement of HRQL be incorporated into routine paediatric rheumatology care [5].

A number of HRQL scales are available for use in children with PRD [6–9]. Some of them have been translated into different languages [10, 11]. However, most of these measures have remained essentially research tools and are not routinely administered in most paediatric rheumatology centres. One of the reasons that may explain why these instruments are uncommonly incorporated in standard clinical care is their length and complexity. To foster regular HRQL assessment in daily practice, there is a need for instruments that are simple, easy to administer and quick.

These considerations have led us to develop a short and simple questionnaire for the assessment of HRQL in routine care of patients with PRD. In this report, we describe the development of the new instrument, the Paediatric Rheumatology Quality of Life (PRQL) Scale, and provide preliminary evidence of its validity in children with juvenile idiopathic arthritis (JIA).

Patients and methods
Development of the PRQL

The PRQL was devised by six paediatric rheumatologists (G.F., C.S.-M., N.R., S.M.M., A.M. and A.R.) with 5 to >20 years of experience in the field and one specialist nurse (D.T.). The panel aimed to devise a short and simple HRQL questionnaire that could be applicable across different PRDs. The instrument was designed to measure the core dimensions of physical, mental and social health delineated by the World Health Organization [12].

Items were derived through: (i) literature review on HRQL in PRD; (ii) analysis of established paediatric HRQL measurements [6–9]; (iii) discussions among members of the study panel; and (iv) semistructured face-to-face interviews of 37 children with different PRDs and their parents regarding their perspective on the ways in which the disease and treatment affected their lives. A total of 389 items were identified. Items were organized into two groups: (i) items related to physical health (PhH) and (ii) items related to psychosocial health (PsH). Items unrelated to these domains were deleted. The study panel then met to consider each potential item. Items were included if they: (i) were general enough to apply to all main PRDs; (ii) were potentially applicable to children of all ages; and (iii) expressed one idea only. At the end of this process, 67 items were left.

After further discussion of the relative importance of each item, an item was retained only when there was an agreement that it should be kept in the questionnaire. This led to a reduction of items to 25, 12 in the physical domain and 13 in the psychosocial domain. Thus, content validity was obtained by the members of the panel. To ensure face validity, the draft questionnaire was shown to eight physicians (four paediatric rheumatologists and four residents in paediatrics), four physiotherapists, two specialist nurses who were not part of the PRQL group and one clinical psychologist, and their opinion on the suitability of the instrument was asked. Although all agreed about the questionnaire, several points were raised regarding definition of items, which were discussed and partially incorporated in the final version. Face and content validity were further tested by asking a convenience sample of 42 children with different PRDs and their parents to complete the draft questionnaire and to criticize or make comments about the design, content, structure and response scale. After these tests, the list was further reduced to 10 items. An item was considered for deletion if: (i) it proved difficult to administer; (ii) it was perceived to lack relevance, to be redundant or to raise some ambiguity in the potential interpretation of the question or the response; and (iii) patients or parents demonstrated difficulty in understanding or completing the item.

A 10-item questionnaire emerged from the analyses. Items were grouped in two subdimensions, each composed by five items: PhH and PsH. For each item or question, a four-point Likert response format [13] was devised. Questions refer the respondent to the previous month. The responses are ‘never’ (score = 0), ‘sometimes’ (score = 1), ‘most of the time’ (score = 2) and ‘all the time’ (score = 3). A ‘not assessable’ column was included in the parent version of the questionnaire to designate questions that cannot be answered because of developmental immaturity. The total score ranges from 0 to 30, with higher scores indicating worse HRQL. A separate score for the PhH and PsH subscales (range 0–15) can be calculated. In case a question is scored as not applicable, the item is given the mean score of the applicable items in the subdimension rounded to the nearest integer. If more than two questions are not answered in each subdomain, the PRQL score cannot be computed. The English translation of the parent proxy report Italian version of the PRQL for ages 2–18 years and child self-report Italian version of the PRQL for ages 7–18 years are presented in supplementary appendices 1 and 2, respectively (available as supplementary data at Rheumatology Online).

Patients and healthy controls

All children aged ≤18 years, seen consecutively at the study units between March 2007 and December 2008 and diagnosed with JIA by the ILAR criteria [14], and their parents (or legal guardians) were enrolled in the study. Patients were excluded if they had another disease or abnormality that could affect HRQL. The control group consisted of healthy children of the same age range recruited at primary and secondary schools in Genova and Pavia and their parents (or legal guardians). Subjects were required to understand Italian. All parents or guardians provided written informed consent to participation in the study. The study was approved by the Institutional Review Board of the Istituto G. Gaslini, Genoa, Italy.
Clinical assessments

The following data were recorded for each patient at study visit: sex, onset age, ILAR category, disease duration and age at visit. Before the visit, a parent of each JIA patient and the patient, if aged ≥7 years, were asked to complete the PRQL independently. A researcher assisted children if they had questions during questionnaire completion. Parents and patients were also asked to rate the child’s overall well-being on a 21-numbered circle visual analogue scale (VAS; 0 = very good; 10 = very poor) [15], the intensity of child’s pain on a 21-numbered circle VAS (0 = no pain; 10 = maximum pain), the presence and duration of morning stiffness and the child’s functional ability by completing the Italian version of the Juvenile Arthritis Functionality Scale (JAFS; 0 = best; 30 = worst) [16]. Parents and children were also asked to rate the current disease status as remission, continued activity or flare and the disease course from previous visit as much improved, slightly improved, stable, slightly worsened or much worsened. Acute phase reactants included ESR and CRP.

The attending physician rated the overall disease activity on a 21-numbered circle VAS (0 = no activity; 10 = maximum activity) [15] and assessed the count of joints with swelling, tenderness or pain on motion, restricted motion and active disease [17]. The physician also rated the current disease status as remission, continued activity or flare, and the disease course from previous visit as much improved, slightly improved, stable, slightly worsened or much worsened.

Healthy schoolchildren and their parents completed the same questionnaires administered to JIA patients and their parents. Children completed the questionnaire during a scheduled break at school. A parent of each child completed the questionnaire at home. Parents’ questionnaires were handed over by children to the school in a closed envelope.

Validation procedures

Validation of the PRQL was conducted following the Outcome Measures in Rheumatoid Arthritis Clinical Trials (OMERACT) filter for outcome measures in rheumatology [18, 19]. Feasibility or practicality was determined by addressing the issues of brevity, simplicity and easy scoring, and from the percentage of missing values [20]. Face and content validity are discussed above.

Criterion validity is a measure of the extent to which values on an instrument agree with those of a gold standard. However, there is no gold standard against which the validity of the PRQL can be tested. For this reason, convergent construct validity was investigated. Construct validity is a form of validation that seeks to examine whether the construct in question, in this case the PRQL, is related to other measurements in a manner consistent with a priori prediction. Given that the PRQL was devised to measure HRQL, we predicted that the correlation with parent’s or patient’s global assessment of child’s well-being and JAFS, which measures a related construct, would be moderate to high. Correlations with pain ratings were also expected to be moderate to high because pain has a major influence on HRQL [21]. Correlations with physician’s global rating and joint counts were predicted to be moderate because, although these measurements assess a different disease construct, active disease and joint impairment may affect markedly the HRQL. Correlations with acute phase reactants were predicted to be low because these parameters have no direct relationship with HRQL. Correlations were expected to be lower for the PsH subscale than for the PhH subscale because PsH is affected by many external factors unrelated to the disease. Correlations were assessed using Spearman’s rank statistics and were considered high if >0.7, moderate if 0.4–0.7 and low if <0.4 [22].

The internal structure of the PRQL was examined using exploratory factor analysis (EFA) with orthogonal varimax rotation [23]. The principal factor method of factor extraction was used and the number of factors to be retained was chosen based on the shape of the scree plot. The EFA generates factor loadings, which are measurements of how strongly the observed variables in the PRQL are associated with its latent factor(s). Internal reliability was assessed using Cronbach’s α coefficient [24]. A value of 0.80 was considered acceptable [25].

To evaluate whether the PRQL can differentiate between patients with varying degrees of disease severity, we compared the median PRQL score and the percentage of subjects with PRQL score = 0 between patients grouped using parents’, patients’ and physicians’ rating of current disease status or disease course from previous visit or level of morning stiffness as external criteria. Comparison of continuous variables was made by Mann–Whitney U-test and comparison of categorical variables was made by chi-square or Fisher’s exact test, as appropriate.

To evaluate test–retest reliability, 35 parents were asked to complete a duplicate copy of the PRQL 24 h after initial administration. This short time frame was chosen to avoid the second assessment being affected by a change in the child’s disease status as a result of a therapeutic intervention (e.g. an IA corticosteroid injection) made at the time of the study visit. Test–retest reliability was assessed by computing the intraclass correlation coefficient (ICC) between the two assessments [26]. An ICC > 0.8 was considered indicative of excellent reliability.

Responsiveness to clinical change over time was assessed by asking parents and patients to complete again the questionnaire during a subsequent visit, after 3–9 months. Responsiveness statistics included the standardized response mean (SRM), calculated as a mean score change divided by s.d. of individuals’ score change. The threshold levels for the SRM were defined as follows: >0.20 = small, >0.50 = moderate and >0.80 = good [27]. The SRM was assessed separately in patients grouped using parents’, patients’ and physicians’ rating of disease course from previous visit as external criteria. The minimal clinically important difference (MCID) in PRQL scores was computed as the mean change in score in patients rated by the parent, the patient or the physician as being slightly improved or slightly worsened from the previous visit.
All statistical tests were two sided; a P-value < 0.05 was considered statistically significant. The statistical packages used were ‘Statistica’ (StatSoft, Tulsa, OK, USA) and Stata release 9.2 (Stata Corporation, College Station, TX, USA).

Results

Patients characteristics

A total of 472 children with JIA, whose main demographic and clinical features are presented in Table 1, were included in the study. A parent of each patient (the mother, whenever possible) completed the PRQL in a total of 1093 visits. In 455 of the 1093 visits, the PRQL was also completed independently by 232 patients aged ≥ 7 years. The scores of the questionnaires completed by parents and patients at study entry are reported in Table 2. Overall, patients tended to rate their HRQL level as better than did their parents. Both parents and patients provided worse ratings for the PhH subscale than for the PsH subscale.

Feasibility and face and content validity

All parents and children reported that the PRQL was simple and easy to understand. Completion and scoring of the PRQL appeared to be quick, both requiring < 5 min. The frequency of missing responses was 1.9 and 2.5% among parents and patients, respectively. The frequency of not assessable responses among parents was 0.4%. Face and content validity are discussed above.

Construct validity

Most correlations for the PRQL were in the predicted range (Table 3). The PhH subscale was correlated at a high level ($r_s > 0.7$) with parent’s assessment of both child’s overall well-being and pain intensity and at moderate level ($r_s = 0.4-0.7$) with JAFS score and tender and active joint counts. Correlations of PhH score with swollen and restricted joint counts and acute-phase reactants were poor ($r_s < 0.4$). Most correlations for the PsH subscale were in the poor range. The construct validity of the PRQL was overall equivalent to that of the Italian version of the Child Health Questionnaire (CHQ) [10] in a convenience sample of 61 JIA patients who had both questionnaires completed by a parent. In the same sample, the physical scale of the PRQL was better correlated with the Italian version of the Childhood Health Assessment Questionnaire (CHAQ) [10] than with the JAFS (results not shown).

Internal structure

EFA with orthogonal varimax rotation on parents’ questionnaires led to the unambiguous identification of two

Table 1 Demographical and clinical features of the 472 study patients at study entry

<table>
<thead>
<tr>
<th>Feature</th>
<th>n (%)</th>
<th>Mean (s.d.)</th>
<th>Median</th>
<th>Lower quartile</th>
<th>Upper quartile</th>
<th>95th percentile</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>108 (22.9)</td>
<td>4.2 (3.3)</td>
<td>2.8</td>
<td>1.7</td>
<td>5.0</td>
<td>10.2</td>
</tr>
<tr>
<td>Female</td>
<td>364 (77.1)</td>
<td>9.0 (4.7)</td>
<td>8.7</td>
<td>4.9</td>
<td>12.2</td>
<td>17.3</td>
</tr>
<tr>
<td>Age at disease onset, years</td>
<td></td>
<td>5.0 (4.0)</td>
<td>3.6</td>
<td>2.7</td>
<td>8.7</td>
<td>13.5</td>
</tr>
<tr>
<td>Age at visit, years</td>
<td></td>
<td>2.5 (3.1)</td>
<td>1</td>
<td>0</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>Physician’s global assessmentb (n = 442), cm</td>
<td>2.4 (2.7)</td>
<td>2.2 (2.8)</td>
<td>0.5</td>
<td>0</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>Parent’s global assessmentb (n = 456), cm</td>
<td>2.0 (2.6)</td>
<td>1.8 (2.3)</td>
<td>0.5</td>
<td>0</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Patient’s global assessment (n = 230), cm</td>
<td>1.8 (2.5)</td>
<td>2.2 (5.0)</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>JAFS score−parentsb (n = 234)</td>
<td></td>
<td>1.9 (3.2)</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>JAFS score−patientsb (n = 234)</td>
<td></td>
<td>1.7 (3.6)</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Swollen joint count (n = 449)</td>
<td></td>
<td>2.3 (5.0)</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Tender joint count (n = 449)</td>
<td></td>
<td>2.2 (5.1)</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Restricted joint count (n = 449)</td>
<td></td>
<td>2.3 (5.0)</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>9</td>
</tr>
<tr>
<td>Active joint count (n = 449)</td>
<td></td>
<td>21.0 (17.3)</td>
<td>15</td>
<td>9</td>
<td>26</td>
<td>58</td>
</tr>
<tr>
<td>ESRd (n = 330), mm/h</td>
<td></td>
<td>1.1 (2.2)</td>
<td>0.46</td>
<td>0.46</td>
<td>0.54</td>
<td>4.9</td>
</tr>
</tbody>
</table>

bOn a 0–10 scale (0 = best; 10 = worst); cOn a 0–30 scale (0 = best; 30 = worst); dnormal < 15 mm/h; dnormal < 0.45 mg/dl.
separate factors: one factor was related to the PhH domain and incorporated items 1–5; the second factor was related to the PsH domain and incorporated items 6–10 (Fig. 1). The straight identification of the two domains was confirmed by the evaluation of factor loadings (results not shown).

Discriminative validity

The PRQL discriminated well between patients classified by the physician, the parent or the patient in different levels of disease severity on the basis of current disease status, disease course from previous visit or level of morning stiffness (Table 4).

**Table 3** Spearman’s correlations between PRQL scores and JIA outcome measures

<table>
<thead>
<tr>
<th>Measure</th>
<th>n</th>
<th>Parent PRQL</th>
<th>Patient PRQL</th>
</tr>
</thead>
<tbody>
<tr>
<td>n</td>
<td></td>
<td>Total score</td>
<td>PhH score</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>PsH score</td>
</tr>
<tr>
<td>Physician’s global assessment</td>
<td>430</td>
<td>0.42a</td>
<td>0.46a</td>
</tr>
<tr>
<td>Parent’s global assessment</td>
<td>444</td>
<td>0.67a</td>
<td>0.70a</td>
</tr>
<tr>
<td>Parent’s pain assessment</td>
<td>445</td>
<td>0.71b</td>
<td>0.74b</td>
</tr>
<tr>
<td>Patient’s global assessment</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Patient’s pain assessment</td>
<td></td>
<td>0.48a</td>
<td></td>
</tr>
<tr>
<td>JAFS score—parents</td>
<td>453</td>
<td>0.61a</td>
<td>0.64a</td>
</tr>
<tr>
<td>JAFS score—patients</td>
<td></td>
<td></td>
<td>0.41a</td>
</tr>
<tr>
<td>Swollen joint count</td>
<td>437</td>
<td>0.35</td>
<td>0.38</td>
</tr>
<tr>
<td>Tender joint count</td>
<td>437</td>
<td>0.44a</td>
<td>0.49a</td>
</tr>
<tr>
<td>Restricted joint count</td>
<td>437</td>
<td>0.37</td>
<td>0.39</td>
</tr>
<tr>
<td>Active joint count</td>
<td>437</td>
<td>0.38</td>
<td>0.42a</td>
</tr>
<tr>
<td>ESR</td>
<td>325</td>
<td>0.38</td>
<td>0.40a</td>
</tr>
<tr>
<td>CRP</td>
<td>331</td>
<td>0.29</td>
<td>0.31</td>
</tr>
</tbody>
</table>

Correlations were tested on questionnaires completed at study entry. See Table 1 for score ranges of outcome measures.

aModerate correlations \(r \geq 0.4, r \leq 0.7\); bhigh correlations \(r > 0.7\).
that the contribution of each item to the questionnaire is well balanced.

**Test–retest reliability**

The ICC for test–retest reliability in 35 parents who completed the PRQL a second time after 24 h was 0.91 for the total score, 0.85 for the PhH subscale and 0.92 for the PsH subscale.

**Responsiveness**

The PRQL was completed at a second visit by 277 parents. The SRMs obtained using a physician’s rating of disease course from a previous visit as external criterion are shown in Table 5. Among patients rated as improved, responsiveness was moderate for the total score and for both subscales. Among patients who were rated as worsened, responsiveness was small for the total score and the PhH subscale and poor for the PsH subscale. As expected, all SRMs values were poor in stable patients.

The MCIDs for the parent PRQL are presented in Table 5. The MCID for the total score ranged between −1.7 and 1.5 in patients rated as slightly improved or slightly worsened by the physician. As expected, all MCIDs in patients rated as stable were close to 0. Responsiveness and MCID could not be assessed for children’s self-reports due to the lack of sufficient longitudinal assessments.

**Healthy children**

The PRQL was completed by 801 parents of healthy schoolchildren, 418 (52.2%) boys and 383 (47.8%) girls, aged 2.2–18.0 years (median 12.1 years) and by 796 healthy schoolchildren, 395 (49.6%) boys and 401 (50.4%) girls, aged 7.3–17.8 years (median 12.5 years).

A joint self- and proxy report of HRQL was obtained for >80% of children aged >7 years. The PRQL scores obtained in healthy children are presented in Table 2. As opposed to what is seen for JIA patients and their parents, healthy children tended to rate their HRQL as worse than did their parents.

**Comparison of PRQL scores between JIA patients and healthy children**

Parents of JIA patients rated their children’s HRQL as more impaired than did parents of healthy children (P < 0.0001). However, the difference depended only on the PhH subscale (P < 0.0001) because the PsH score was comparable (P = 0.34). JIA patients self-reported their overall HRQL as worse than did healthy children (P = 0.03). Again, this difference was due to the greater

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**FIG. 1** Factor analysis with orthogonal varimax rotation leading to unambiguous identification of two separate factors: one factor was related to PhH domain and incorporated items 1–5, the second factor was related to PsH domain and incorporated items 6–10.

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**Table 4** Discriminative ability of the PRQL by level of disease severity as rated by physicians, parents and patients

<table>
<thead>
<tr>
<th>Assessment of disease status at study entry*</th>
<th>Parent’s ratings</th>
<th>Parent’s ratings</th>
<th>Patient’s ratings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Parent PRQL total score</td>
<td>Parent PRQL total score</td>
<td>Parent PRQL total score</td>
</tr>
<tr>
<td></td>
<td>Median (IQR) n (%) with score = 0</td>
<td>Median (IQR) n (%) with score = 0</td>
<td>Median (IQR) n (%) with score = 0</td>
</tr>
<tr>
<td>Remission</td>
<td>2 (0–3) 106 (32.1)</td>
<td>1 (0–3) 105 (34.2)</td>
<td>1 (0–3) 49 (33.3)</td>
</tr>
<tr>
<td>Continued activity</td>
<td>6 (3–9) 6 (5.4)</td>
<td>5 (3–9) 12 (6.8)</td>
<td>5 (2–7) 4 (6.3)</td>
</tr>
<tr>
<td>Flare</td>
<td>8 (5–11) 0 (0)</td>
<td>4 (2–7) 11 (6.5)</td>
<td>4 (2–7) 3 (4.2)</td>
</tr>
<tr>
<td>Assessment of disease outcome from previous visit**</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Improved</td>
<td>1 (0–3) 84 (35.6)</td>
<td>2 (0–4) 85 (28.3)</td>
<td>2 (1–4) 41 (24.8)</td>
</tr>
<tr>
<td>Stable</td>
<td>4 (2–8) 25 (12.8)</td>
<td>3 (1–6) 37 (17)</td>
<td>2.5 (1–6) 13 (17.1)</td>
</tr>
<tr>
<td>Worsened</td>
<td>4 (2–9) 5 (7.1)</td>
<td>5 (2–9) 6 (5.1)</td>
<td>4 (2–6) 1 (1.3)</td>
</tr>
<tr>
<td>Assessment of morning stiffness*</td>
<td>–</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>2 (0–4) 120 (28.1)</td>
<td>2 (0–4) 50 (26.5)</td>
<td></td>
</tr>
<tr>
<td>≤30 min</td>
<td>–</td>
<td>5 (3–9) 10 (6.9)</td>
<td>3 (2–7) 5 (6.2)</td>
</tr>
<tr>
<td>&gt;30 min</td>
<td>–</td>
<td>8 (5–12) 0 (0)</td>
<td>5.5 (4–7) 0 (0)</td>
</tr>
</tbody>
</table>

This analysis was made only for parent-reported PRQL. *P < 0.0001 for all comparisons; **P < 0.0001 for all comparisons, except for patient’s rating of disease outcome from previous visit (P = 0.003).
PhH score provided by patients ($P = 0.0002$), whereas the PsH score was higher (worse) in healthy children ($P < 0.0001$). These differences were not related to disparities in sex because PRQL scores in boys and girls were comparable among either patients or healthy children (results not shown). Stratification of patients and healthy children by age (<10, 10–13 and >13 years) showed that the difference for the PsH subscale was much more pronounced in children >13 years for both proxy- and self-reported HRQL (results not shown).

To scrutinize the ability of the PRQL to discriminate between sick and healthy children, we compared scores obtained in healthy children with scores obtained in JIA patients stratified by severity of joint disease (no active joints, 1–4 active joints and ≥5 active joints). As expected, the PhH score of patients with no active joints was comparable to that of healthy children, whereas patients with ≥1 active joints had a PhH score greater than that of healthy children ($P < 0.01$). An opposite phenomenon was seen for the PsH score, with patients with no active joints or with 1–4 active joints providing better scores than healthy children, whereas the scores of patients with ≥5 active joints were comparable with those of healthy children. Patients with ≥5 active joints had a worse PsH score than those with 1 active joint ($P < 0.01$; results not shown).

**Discussion**

We have described the development of a new measurement of HRQL for patients with PRD and its preliminary validation in children with JIA. The PRQL is short and simple, and is quick, taking <5 min to complete and score, which makes it practical for use in standard clinical care. It is proposed for use as both proxy report and patient self-report, with the suggested age range of 7–18 years for use as self-report. The Italian version of the questionnaire was found to be feasible and to possess face and content validity, good construct validity, satisfactory reliability and internal structure and consistency, strong discriminative validity and fair responsiveness to clinically important change over time in a large cohort of patients with JIA. By documenting these key measurement properties, we have demonstrated that the PRQL is a valid tool for the assessment of HRQL in this patient population and is, therefore, applicable in both clinical and research settings.

As recently observed in a large multinational study [28], we found that both proxy- and self-reported HRQL were more impaired in JIA patients than in healthy children, with PhH being most involved. Unexpectedly, however, the level of psychosocial well-being of JIA patients was comparable to (for parents’ proxy reports) or even better than (for children’s self-reports) that of healthy children. This phenomenon may depend on most of the JIA patients attending our clinics for follow-up visits having well-controlled disease with little or no disease activity or disability. Children with JIA suffer in the active phase of their disease a considerable burden of symptoms, namely pain and stiffness, which affects many aspects of their life. For these children, disease improvement represents a key priority. It is, therefore, conceivable that relief of symptoms leads to a marked enhancement in their mental and social health. An indirect confirmation of this hypothesis came from the observation that the psychosocial PRQL score was significantly worse in JIA patients with more extended arthritis than in those with monoarticular disease.

Healthy children have different priorities, particularly regarding comparison of their look and abilities with those of their peers. Importantly, the poorer PsH seen in healthy children concerned almost exclusively the adolescent age group. Adolescents are medically, developmentally and psychologically distinct from children and from adults [29]. Furthermore, adolescence is known to be a critical age that entails considerable emotional and psychological distress [30]. To our knowledge, our study is the first to show that mental and social functioning of healthy adolescents is comparable to or even worse than that of adolescents with JIA. This observation deserves further exploration in different populations.

We cannot exclude the possibility that these findings could depend on a poor ability of the PRQL to capture the psychosocial components of HRQL. However, similar
results were recently reported by Trapanotto et al. [11], who administered an established and more detailed HRQL tool, the PedsQL 4.0 Generic Core Scales, to an Italian sample of children with rheumatic diseases and healthy children. They found a significant difference between children with rheumatic diseases and healthy children only in emotional functioning for both self- and proxy reports, with children with rheumatic diseases having, however, significantly higher scores, meaning better HRQL. For all the other dimensions of HRQL, either physical or psychosocial, the difference between sick and healthy children was not significant. To explain this phenomenon, the authors hypothesized that at the time of the assessment most patients had achieved disease remission. As a result, they were experiencing only little or no limitation of their daily activities, and for this reason, the global level of their quality of life was good.

Notably, the use of another more comprehensive HRQL tool, the Juvenile Arthritis Quality of Life Questionnaire (JAQQ), in adolescents demonstrated shortcomings. Shaw et al. [31] reported that as many as one-fifth (21.8%) of 308 adolescents with JIA failed to answer all 78 items included in the questionnaire. They concluded that as a consequence of the comprehensiveness of the instrument, there is a danger that clinically important information can be lost.

We found that JIA patients tended to rate their HRQL level better than did their parents. A similar observation was reported for the assessment of pain and overall well-being [21]. This suggests that either children may cope with their disease better than realized by parents or parents tend to be overly solicitous of their children’s health problems. An opposite trend was seen for healthy children, however, with parents’ proxy reports being more optimistic than children’s self-reports. As above, this was most common among adolescents. In a recent study, adolescents were found to be less optimistic than their parents with respect to mental health, well-being, general health and impact of health on family activities [32]. A wide variation in agreement between adolescents with JIA and their parents about health assessments, including HRQL, was reported [33]. Overall, these studies emphasize the importance of considering both parents’ and children’s perspectives in the assessment of HRQL.

Our study should be viewed in the light of certain limitations. Although we present the English translation of the questionnaires (Appendices 1 and 2, available as supplementary data at Rheumatology Online), the instrument was tested in Italian patients and healthy children. It is possible that children and their parents elsewhere might respond differently to the PRQL questionnaire due to cultural and language differences. The low level of disease activity and disability in most of our patients may have limited the generalizability of our study. In particular, it might have hampered the assessment of responsiveness to clinical change over time, especially for worsening of disease. However, our patients represent a consecutive sampling of our clinic population and are likely representative of the patients seen in most tertiary paediatric rheumatology centres. Nevertheless, our results should be confirmed at other sites and in different patient settings before the new questionnaire is widely adopted. We should recognize that healthy controls were, on average, older than JIA patients, which could add to the explanation of the lack of responsiveness of the PsH domain of the questionnaire. The PRQL addresses only the dimensions of physical, mental and social health, and lacks assessment of wider, family-related HRQL. We acknowledge that our work owes a great deal to previous work on development and validation of HRQL questionnaires for PRD and that some of the items in the tool are the same as or very similar to those in the JAQQ [6], the PedsQL [7] and the CHQ [8].

In conclusion, we have developed a new short and simple measure for the assessment of HRQL in patients with PRD, which is primarily proposed for use in standard clinical care. This instrument, which was validated in children with JIA in its Italian version, should be further tested in patients with other PRDs and in different cultural environments.

Rheumatology key messages

- Evaluation of HRQL is fundamental in children with rheumatic diseases.
- Existing HRQL measures are rarely used in daily practice due to their complexity.
- The PRQL is a simple questionnaire suitable for the assessment of HRQL in routine care.

Disclosure statement: The authors have declared no conflicts of interest.

Supplementary data

Supplementary data are available at Rheumatology Online.

References