

# Health-Related Quality of Life of Children and Adolescents with Juvenile Idiopathic Arthritis in Western Saudi Arabia

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## Abstract

**Objectives:** To evaluate the effect of Juvenile Idiopathic Rheumatoid Arthritis (JIA) on the health-related quality of life (HRQOL) in Saudi children. **Methods:** A cross-sectional study was conducted in a tertiary hospital in Jeddah, Saudi Arabia to evaluate the HRQOL of children aged  $\leq 18$  years who had JIA using the childhood health assessment questionnaire modified for Arab children (CHAQ-MAC). Such questionnaire investigates 34 activities of daily life (ADL) classified into 8 life domains. Children or their parents were invited for face-to-face interview, and a phone interview was done for patients who missed their appointments during the period between February and July 2017. A statistical model was used to calculate a total CHAQ.MAC score (range = 0 - 33; Cronbach's alpha = 0.966); with higher values indicating poorer HRQOL. **Results:** Of a total of 44 children (male ratio = 0.63; mean  $\pm$  SD age =  $9.95 \pm 5.44$ ), Systemic-onset JIA was the most frequent type (27.3%), followed by polyarticular (15.9%) and oligoarticular (13.6%). Pain was reported among 43.2% (frequently in the knee, in 27.3%) whereas morning stiffness was reported in 20.5%. The mean CHAQ.MAC score = 2.89 (75<sup>th</sup> centile = 3.00). With respect of ADLs, up to 22.7% of the children complained of difficulty; and 31.8% reported a difficulty in at least one of the 34 investigated ADLs. With respect of the life domain, children reported difficulties for activities (27.3%), dressing & grooming and hygiene (13.6%), and eating (6.82%). According to the life domain, 4.5% to 13.6% of the children needed help to execute the related ADLs and up to 9.1% used aids or devices. Poor HRQOL was associated with articular pain ( $p = 0.003$ ) and specific medication ( $p = 0.043$ ). **Conclusion:** Children with arthralgia and those on specific treatment are at higher risk of impaired QOL, which emphasizes the need for systematic screening for treatment adverse effects and joint pain and implementation of efficient management to improve HRQOL.

## Keywords

Juvenile Idiopathic Rheumatoid Arthritis, JIA, Quality of Life, Disability, Activities of Daily Life

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## 1. Introduction

Being the most commonly-reported rheumatic disease in childhood, juvenile idiopathic arthritis (JIA) represents a major health problem as a set of heterogeneous conditions that have been formerly considered as separate clinical entities [1]. The first description of a chronic form of childhood arthritis has been established in 1896 by the English paediatrician George Frederick Still [2], with a subsequent investigation of juvenile rheumatism in Canada in 1946 and framing of childhood arthritis by the American College of Rheumatology, namely juvenile rheumatoid arthritis [3].

The International League of Associations for Rheumatology (ILAR) was adopted for our study which will be discussed later in the paper including the description of symptoms related to age [4] [5]. In general, the prevalence of JIA shows a remarkable variation across different countries due to the diversity of frequency estimates and the lack of identifiable uniform classification methods worldwide. Evidence has shown that disease incidence ranges between 2 and 22 per 100,000 populations, while prevalence estimates indicate figures between 7 and 150 per 100,000 [6] [7] [8]. In the United States, Peterson *et al.* [9] have found a significant reduction of JIA incidence per 100,000 population from 15 to 7.8 during the period from 1960-1969 to 1980-1993, respectively, particularly for the systemic-onset type. Seemingly, JIA is more prevalent in northern Europe when compared to other regions [10]. However, an interesting finding in an Australian study revealed that disease prevalence reached up to 400/100,000 [11]. Ethnicity may have also a significant impact on the distribution of JIA subcategories given the scarcity of anti-nuclear antibodies (ANA)-positive JIA in India [12] [13]. Few epidemiological investigations have been conducted in the Middle East, with a prevalence rate of 3.43 and 20 in Egypt and Oman, respectively [14] [15]. To the best of our knowledge, there are no prevalence estimates of JIA in Saudi Arabia based on reliable nationwide or multicentre-based studies.

Given the high frequency of JIA as a chronic illness during childhood, it is plausible to assess disease outcomes over long periods. Nonetheless, the main complications differ according to disease subtype. For instance, the most important unfavorable outcome in children with chronic oligoarthritis is chronic uveitis, which is more pronounced in ANA-positive conditions [16]. Additionally, some patients with enthesitis-related JIA would develop ankylosing spondylitis, cerebrovascular, and cardiopulmonary complications, which would ultimately shorten the patients' life expectancy. Amyloidosis is also a frequent complication with the persistent inflammatory reactions that could be seen in systemic JIA

[17].

Importantly, the involvement of physical activity, emotional aspects, and sociological well-being of children and adolescents in JIA might impact the health-related quality of life (HRQOL). The latter concept was originally utilized in the early 1990s to express not only the subjective evaluation implied by the laboratory and imaging investigations but also the personal assessment and analysis of treatment outcomes [18]. For adequate assessment, multiple dimensions are essentially utilized and the assessment would also include several items within each dimension. In addition, HRQOL can be regarded also as an important indicator of patients' psychological health [19] [20]. As such, assessment of the functional status is crucial to evaluate disease-attributable disability and pain and their impact on the QOL. This could be performed via several useful tools, such as the childhood health assessment questionnaire. Collectively, in light of the relative lack of knowledge regarding JIA consequences in Saudi Arabia, we conducted this study to assess the QOL relying on investigating the activities of daily life (ADL) using the childhood health assessment questionnaire modified for Arab children (CHAQ-MAC) in children with different subcategories of JIA.

## 2. Materials and Methods

### 2.1. Criteria of JIA

The currently-identified classification of JIA, which has been proposed in 2001 by the International League of Associations for Rheumatology (ILAR), includes seven subgroups based on the predominant laboratory properties, including the following patterns of arthritis: systemic involvement, oligoarticular, polyarticular (with a positive or negative rheumatoid factor), psoriatic, enthesitis-related, and an unclassified form. The clinical diagnosis of JIA entails the existence of a swelling or movement limitation of a joint associated with pain, heat, or tenderness without apparent aetiologies for arthritis. Such symptoms occur in children less than 16 years of age and should persist for at least 6 weeks. Basically the patient selection was chosen according to the (ILAR) criteria and they were mainly of JIA patients and patients with Psoriasis were not included, as we don't have a good number to be enrolled in the study.

### 2.2. Design and Setting

This is a cross-sectional study that was carried out among children aged  $\leq 18$  years, following for JIA at the Paediatric Rheumatology outpatient clinic, inpatient ward and daycare unit in King Abdul-Aziz University Hospital (KAUH), Jeddah, Saudi Arabia, in the period February 2017-July 2017. The study protocol was approved by the KAUH institutional review board.

### 2.3. The Questionnaire

Assessment of quality of life used the childhood health assessment questionnaire-modified for Arab children (CHAQ-MAC) which was first adopted and

used by *Madi et al.* [21] which is a tool developed to assess the health-related quality of life (QoL) among children afflicted with juvenile rheumatoid arthritis. The tool was adapted and validated for Arab populations, showing adequate feasibility and reliability features along with strong correlation with disability index. The CHAQ-MAC assesses the child's level of difficulty regarding 34 activities of daily life (ADL) classified into 8 life domains: dressing & grooming (4 items), arising (2 items), eating (3 items), walking (2 items), hygiene (6 items), reach (4 items), grip (6 items), and activities (7 items). Each of the items is a 4-point likert-type scale ranging from "unable to do" to "no difficulty/without aid". Further, the questionnaire investigates the use of any aids or devices, along with assessment of overall health status and pain score using a 0 - 10 visual analogue scale (VAS).

#### **2.4. Scoring System**

In the present study, a modified scoring system was used, based on a binomial answering method, where existence of difficulty was scored as 1 and absence of difficulty was scored as 0, for each of the activity items. This resulted in a total CHAQ-MAC score ranging from 0 - 34, with higher values indicating poorer QoL. Cases with non-applicable answers were scored as 0, assuming absence of difficulty in the given item.

#### **2.5. Data Collection Procedure**

To administer the questionnaire, children and their parents were invited for a face-to-face interview. According to the respondent suitability, the questionnaire was administered in Arabic or English language. Participants who missed their appointment were re-contacted and the questionnaire was administered via phone interview.

#### **2.6. Statistical Methods**

Statistical analysis was performed with the Statistical Package for Social Sciences version 21.0 for Windows (SPSS Inc., Chicago, IL, USA). The questionnaire reliability was tested by calculation of the Cronbach's alpha. Descriptive statistics were used to present demographic and clinical characteristics of the patients; categorical variables are presented as frequency and percentage, while continuous variables are presented as mean  $\pm$  standard deviation (SD). Nonparametric tests were used to analyze factors associated with CHAQ-MAC score; Mann-Whitney U test was for binomial variables and Kruskal-Wallis test for multinomial ones, and results are presented as mean CHAQ-MAC score with 75<sup>th</sup> centile, as the median was null for majority items. A p value of <0.05 was considered to reject the null hypothesis.

### **3. Results**

#### **3.1. Participants' Characteristics**

Forty-four children with JIA were included; male ratio = 0.63; mean  $\pm$  SD age =

9.95 ± 5.44 years. Systemic-onset JIA was the most frequent type (27.3%), followed by polyarticular (15.9%) and oligoarticular (13.6%); and 47.7% of the patients were on treatment. Pain was reported among 43.2%, with knee being the most frequent localization (27.3%); whereas morning stiffness was reported in 20.5% (**Table 1**).

### 3.2. Reliability Testing of the CHAQ-MAC Questionnaire

In the present study population, the modified version of the questionnaire (CHAQ-MAC) showed excellent reliability with Cronbach's alpha = 0.966 (33 items, one item excluded for null variance). With respect of the subscale, Cronbach's alpha values varied between 0.693 to 0.900 (Supplemental **Table 2**).

**Table 1.** Demographic and clinical characteristics and general health status of children with JIA.

Parameter	Category	Frequency	Percentage
Age (years)	Mean, SD (range = 0 - 18)	9.95	5.44
Gender	Male	17	38.6
	Female	27	61.4
	SJIA	12	27.3
Type of onset	Polyarticular	7	15.9
	Oligoarticular	6	13.6
	Psoriatic	2	4.5
	Do not know	17	38.6
Taking medication	Yes	21	47.7
	No	21	47.7
	Do not know	2	4.5
Pain	Yes	19	43.2
	No	22	50.0
	Do not know	3	6.8
Past week pain	Knee	12	27.3
	Wrist	5	11.4
	Shoulder	1	2.3
	Neck	0	0.0
	Do not know	1	2.3
Pain intensity (VAS score)	None	25	56.8
	Mean, SD	2.78	3.49
Health status (VAS score)	Mean, SD	2.27	3.55
	Yes	9	20.5
Morning stiffness	No	34	77.3
	Do not know	1	2.3

SD: Standard deviation; SJIA: systemic-onset juvenile idiopathic arthritis; VAS: visual analog scale.

**Table 2.** (Supplemental): Reliability analysis of modified CHAQ.MAC and subscales.

Questionnaire subscale	No. of items	Cronbach's alpha
CHAQ.MAC (modified version)	33	0.966
<b>Subscales</b>		
Dressing & grooming	4	0.900
Arising	2	0.873
Eating	3	0.738
Walking	2	0.841
Hygiene	5*	0.881
Reach	4	0.740
Grip	6	0.693
Activities	7	0.879

\*"Use Arabic style toilet" item was excluded as non-applicable for majority of the participants.

### 3.3. Quality of Life Assessment

Results of QoL assessment are presented in detail in **Table 3** and **Table 4**; and a summary is presented in the present paragraph. Mean CHAQ.CAM score was 2.89 (75th centile = 3.00). With respect of ADLs, up to 22.7% of the children complained of difficulty; and 31.8% reported a difficulty in at least one of the 34 investigated ADLs. With respect of the life domain, the percentage of children who reported difficulties varied between 6.82% (mainly cutting meat) for eating and 27.3% (lifting cups) for activities; while both dressing/grooming 9.1% (dressing), and regards to hygiene 4.5% (taking bath and washing hair was difficult for most) Furthermore, 13.6% of the children needed help to execute the related ADLs (mainly arising/gripping and opening), and up to 9.1% used aids or devices (bath bar). Consequently, 6.8% to 13.6% of JIA children need help to achieve ADLs and aids and or devices are used in 4.5% to 9.1% depending on the specific ADL.

### 3.4. Factors of Poor Quality of Life

Children under specific medication had poorer QoL as indicated by mean (75<sup>th</sup> centile) CHAQ.CAM score = 5.38 (8.66), which was higher than their counterparts (0.52 [1.21]), and the difference was statistically significant ( $p = 0.043$ ). Similarly, presence of articular pain was associated with poorer QoL (6.00 [14.00] versus 0.59 [0.00]) by comparison to absence of pain ( $p = 0.003$ ) (**Figure 1**). On the other hand, no significant association was found between QoL and age ( $p = 0.353$ ), gender ( $p = 0.080$ ) or type of onset ( $p = 0.400$ ) (**Table 5**).

## 4. Discussion

The quality of life (QOL) of patients with chronic disease is variably affected and it comprises a multidimensional construct, including at least the social, physical, and psychological functionality. Disease outcomes and the impact of its targeted

**Table 3.** Difficulty assessment in activities of daily life among children with JIA.

Domain/Activity	No difficulty		Some difficulty		Much difficulty		Unable to do		Not applicable	
	F.	%	F.	%	F.	%	F.	%	F.	%
<i>Dressing &amp; grooming</i>										
Dress	39	88.6	4	9.1	0	0.0	0	0.0	1	2.3
Shampoo	39	88.6	3	6.8	1	2.3	0	0.0	1	2.3
Remove socks	40	90.9	3	6.8	0	0.0	0	0.0	1	2.3
Cut fingernails	38	86.4	3	6.8	0	0.0	0	0.0	3	6.8
<i>Arising</i>										
Stand up	38	86.4	3	6.8	2	4.5	0	0.0	1	2.3
Get in & out bed	38	86.4	3	6.8	2	4.5	0	0.0	1	2.3
<i>Eating</i>										
Cut meat	39	88.6	3	6.82	0	0.0	0	0.0	2	4.5
Feed with hand	40	90.9	2	4.5	0	0.0	0	0.0	2	4.5
Lift a cup	42	95.5	1	2.3	0	0.0	0	0.0	1	2.3
<i>Walking</i>										
Walk outdoors	39	88.6	4	9.1	0	0.0	0	0.00	1	2.3
Climb up	39	88.6	1	2.3	3	6.8	0	0.0	1	2.3
<i>Hygiene</i>										
Wash body	39	88.6	2	4.5	2	4.5	0	0.0	1	2.3
Taking bath	39	88.6	3	6.8	1	2.3	0	0.0	1	2.3
Brush teeth	42	95.5	1	2.3	0	0.0	0	0.0	1	2.3
Brush hair	39	88.6	3	6.8	0	0.0	0	0.0	2	4.5
American toilet	38	86.4	2	4.5	1	2.3	2	4.5	1	2.3
Arabic toilet	4	9.1	0	0.0	0	0.0	0	0.0	40	90.9
<i>Reach</i>										
Heavy object	36	81.8	2	4.5	3	6.8	1	2.3	2	4.5
Bend down	37	84.1	4	9.1	1	2.3	0	0.0	2	4.5
Pull on	40	90.9	3	6.8	0	0.0	0	0.0	1	2.3
Look back	41	93.2	2	4.5	0	0.0	0	0.0	1	2.3
<i>Grip</i>										
Write	41	93.2	2	4.5	0	0.0	0	0.0	1	2.3
Open car	41	93.2	1	2.3	0	0.0	0	0.0	1	2.3
Open jar	36	81.8	2	4.5	1	2.3	2	4.5	3	6.8
Turn faucets	38	86.4	1	2.3	2	4.5	1	2.3	2	4.5
Open can	41	93.2	0	0.0	0	0.0	0	0.0	3	6.8
Open door	38	86.4	3	6.8	1	2.3	1	2.3	1	2.3
<i>Activities</i>										

**Continued**

Get in/out car	40	90.9	3	6.8	0	0.0	0	0.0	1	2.3
Ride bike	35	79.5	2	4.5	1	2.3	1	2.3	5	11.4
Run and play	38	86.4	2	4.5	1	2.3	1	2.3	2	4.5
Households duties	38	86.4	3	6.8	1	2.3	0	0.0	2	4.5
Cross sitting	33	75.0	9	20.5	0	0.0	1	2.3	1	2.3
Prayer position	34	77.3	6	13.6	1	2.3	1	2.3	2	4.5
Errands & shop	36	81.8	4	9.1	2	4.5	0	0.0	2	4.5

JIA: Juvenile idiopathic arthritis. F. Frequency; % percentage.

**Table 4.** Help needed and aids used among children with JIA.

Parameter	Activity/aid	Frequency	Percentage
Help needed for...	Eating	3	6.82
	Dressing and grooming	5	11.4
	Walking	3	6.8
	Arising	6	13.6
	Hygiene	2	4.5
	Gripping & opening	6	13.6
	Reach	3	6.8
	Errands and chores	5	11.4
	Cane	2	4.5
	Walker	2	4.5
Aids and devices used	Crutches	2	4.5
	Wheelchair	2	4.5
	Built up utensil	2	4.5
	Special chair	2	4.5
	Device for dressing	2	4.5
	Raised toilet seat	4	9.1
	Bathtub seat	2	4.5
	Jar opener	2	4.5
	Bathtub bar	4	9.1
	Bathroom Appliances	3	6.8
Reach Appliances	1	2.3	

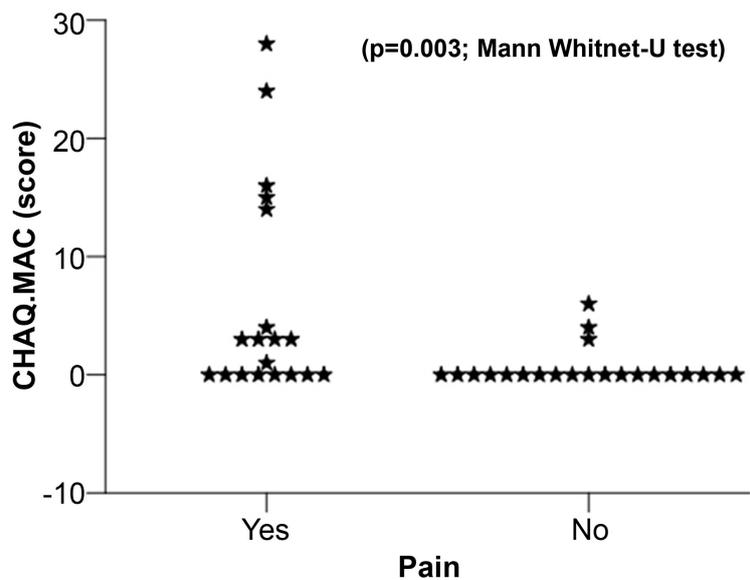
**Table 5.** Demographic and clinical factors associated with quality of life among children with JIA (nonparametric tests).

Parameter	Category	Quality of life (CHAQ.MAC score)		p-value
		Mean	75 <sup>th</sup> centile	
Age (years)	0 - 5	1.00	3.00	0.353
	>5 - 10	5.58	11.50	
	>10 - 15	1.79	0.75	
	>15	3.43	0.00	

## Continued

Gender	Male	0.59	0.00	0.080
	Female	4.33	4.00	
	SJIA	1.25	0.00	
Type of onset	Polyarticular	4.43	3.00	0.400
	Oligoarticular	3.00	6.50	
	Psoriatic	1.50	0.00	
	Do not know	3.53	3.50	
Taking medication	Yes	5.38	8.66	0.043*
	No	0.52	1.21	
Pain	Present	6.00	14.00	0.003*
	Absent	0.59	0.00	
Past week pain	Knee	0.58	0.00	0.484
	Wrist	3.60	9.00	
	None	2.84	3.00	
Pain severity (VAS score)	Low (0)	1.00	3.00	0.345
	Moderate (1 - 5)	5.86	14.00	
	Severe (>5)	6.80	15.50	
Health status (VAS score)	Good (0)	0.88	2.25	0.138
	Moderate (1 - 5)	10.80	20.00	
	Bad (>5)	3.60	9.00	
Morning stiffness	Yes	9.44	20.00	0.061
	No	1.15	0.25	

SD: Standard deviation; SJIA: systemic-onset juvenile idiopathic arthritis; VAS: visual analogous scale; \*statistically significant result; tests used: Manny Whitney-U test for binomial variables and Kruskal Wallis test for multinominal variables.



**Figure 1.** Association of pain with quality of life among children with JIA. The greater the score the worse the quality of life.

interventions are reflected as HRQOL and thus such concept can be deemed a primary outcome and a fundamental determinant of the therapeutic benefits [22]. However, some researchers may use the HRQOL as a secondary outcome to generate hypotheses. In the context of JIA, both pain and medication-attributable adverse effects have been significantly associated with worsening of the HRQOL in the present study, a matter which can be considered in future intervention studies. We demonstrated also that the HRQOL is affected in approximately one-third of patients and, to a less extent, they required assistance during performing their ADLs.

The mean CHAQ.MAC score in the present study was 2.89 and the majority of children scored  $\leq 5$  (out of 33), indicating favorable outcomes despite the existence of systemic symptoms in 27.3% of patients. Amine *et al.* [23] have conducted a CHAQ-based study and showed a higher overall mean score (0.84), where a score of 3 indicated maximum disability. The use in our study of a different scale is justified by the very low scores; as the use of a 0 - 3 scale would produce unintelligible values. In the previously mentioned study by Amine *et al.* [23], the HRQOL decreased remarkably with increased disability index as indicated by higher CHAQ scores. The CHAQ was also a beneficial tool to discriminate between JIA patients and the healthy peers in a case-control study conducted in Egypt, showing a significant difference between both groups in terms of the disability index ( $1.1 \pm 0.8$  vs  $0.1 \pm 0.2$ , respectively,  $p < 0.01$ ) [24]. Additionally, cross-sectional studies relying on the CHAQ score showed that motor functions were generally worse in patients with polyarticular and systemic JIA when compared to the oligoarticular subtype ( $p < 0.05$ ) even if the symptoms persisted for long times [24] [25] [26]. In the current study, we showed a relative increase in the mean CHAQ scores in the systemic and polyarticular types even though the difference with oligoarticular JIA was insignificant.

Despite the evident impact of disability index, the effect of functional outcome in JIA on HRQOL seems to be less apparent. In the present study, only  $\leq 13\%$  of patients required assistance to perform ADLs and less than 9% used aids and devices. The use of early aggressive therapy, including the intra-articular injection of steroids and methotrexate, in JIA patients usually yields an improved outcome and their physical abilities are frequently preserved. Our results are consistent with those reported by Ravelli [27] who demonstrated that only 10% of children with JIA would have severe disability. Marked functional disability might develop in 12% of patients within the first 5 years and in 50% following 16 years of disease onset [28]. Besides, performing household activities, mobility, and physical activity are mildly impaired in children with functional independence. Poor physical activity would possibly occur in patients with positive rheumatoid factor (RF), polyarticular JIA, radiological damage, and pain in the wrist and hip rather than hand joints [29].

The essence of altering the QOL in children and adolescents with chronic arthritis is experiencing pain. Painful episodes were reported in approximately half of patients under study and it was associated with poorer QOL. Previous

reports have shown that JIA symptoms affected the sociological, emotional, and physical aspects in approximately 1 in each 1000 children aged less than 16 years [30]. JIA patients suffer from joint pain, morning stiffness, and fatigue, which can be associated with dramatic changes in the HRQOL. Indeed, Sawyer *et al.* [31] emphasized the significant and independent relationship between pain and different pain coping strategies with various HRQOL-related domains. The higher levels of experienced pain the greater impairment of physical, social, and emotional functioning in JIA patients as reported by both the patients and their parents. Likewise, another study [23] among Moroccan children and adolescents has revealed the significant correlation between pain, joint swelling, and high erythrocyte sedimentation rates as indicators of disease activity and poor HRQOL ( $P < 0.0001$ ). In contrast to our results, however, the involvement of multiple joints in the polyarticular and systemic subtypes was also associated with increased pain and poorer HRQOL. In addition, the existence of pain, but not pain severity, was the impactful factor of altering the HRQOL. It is worth noting that pain worsening may be regarded a crucial independent risk factor of depression, which increases the overall burden and deteriorate HRQOL [32].

From another perspective, treatment adverse effects had a major effect on the QOL of JIA patients although evidence from the literature demonstrates relatively contrasting findings. JIA treatment comprised originally the use of non-steroidal anti-inflammatory drugs (NSAIDs) and corticosteroids to relive pain and reduce inflammation but they can be used currently as adjunctive therapies. Intra-articular corticosteroid injection is a safe procedure that provides local and long-lasting effects and it can replace systemic therapies in the persistent oligoarticular JIA [33]. Using methotrexate, a traditional disease-modifying anti-rheumatic drug, is common in JIA via the oral and subcutaneous routes, but it may be associated with gastrointestinal adverse effects and impaired hepatic and blood count parameters [34]. However, it seems that the effects of these symptoms are not significant given the improvement of HRQOL components by methotrexate therapy, especially the physical domain, as revealed by the Paediatric Rheumatology International Trials Organization (PRINTO) [35]. Sulfasalazine can be also used but the current recommendations indicate its use in enthesitis-related RIA since it causes severe toxicities in adult-onset Still disease and therefore should be excluded in the related JIA conditions [36]. Importantly, the use of biologic therapies, including tumour necrosis factor inhibitors (TNFi) and interleukin-1 inhibitors, have achieved a tremendous progress during the past decade in polyarticular, psoriatic, and enthesitis-related JIA [37]. Etanercept, one of the TNFi, has been shown to exert beneficial actions on multiple domains of HRQOL, including the emotional and functional domains [38].

Emphasising the role of improving patients' activities and independence should be not only limited to young-aged patients but also to the long-term effects in adolescents and adults. Reducing physical impairment and disability via introducing an aggressive treatment is crucial to enhance independence and

support self-worth and self-esteem. These targets should be achieved without considering the existence of functional disability during the transition from childhood to adulthood. As such, adequate vocational planning and improving the personal skills should be established in JIA patients for future prospects.

This is the first study in Saudi Arabia which investigated the HRQOL in JIA children and adolescents. We provided an outline about disease onset in a large tertiary hospital that represents a considerable proportion of the population despite the apparent small sample size. The used questionnaire showed an overall excellent reliability through high internal consistency scores for the main questionnaire and activity subscales (Cronbach's alpha > 0.7). However, some limitations are involved, including the potential variation in disease classification with other studies and the lack of data about the prescribed treatments. Finally, recruiting patients from a tertiary medical centre might induce biased outcomes toward severe JIA patterns.

## 5. Conclusion

Information on the impact of JIA can be utilized clinically to make the provided health services more patient-centered. The dual fundamental outcomes of HRQOL assessment, namely the effect of a disease and the impact of medical interventions, were evident in the present study by the significant association between lower level of QOL and articular pain and receiving a specific medication, respectively. Such findings highlight the need for continuous screening for joint pain and treatment-related adverse effects during management. A lot of specialties could be included in the patient's care, including psychologists and social workers to improve the overall HRQOL in JIA. Studies investigating the QOL of adult patients with JIA and its impact on the educational attainment as well as employment rates are warranted.

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## Data Availability

All data generated or analyzed during this study are included in this article. Spreadsheet (or dataset, or any other files) is available upon request from authors, original modified questionnaire will be sent by e-mail along with the manuscript.

## Funding Statement

This study required no funding as it is based on simple data collection via inter-

viewing parents and patients during clinic visits, admissions and day care visits.

## Supplementary Materials

All data are included in the article.

## Conflicts of Interest

No conflicts of interest are declared by the authors.

## References

- [1] Giancane, G., Consolaro, A., Lanni, S., *et al.* (2016) Juvenile Idiopathic Arthritis: Diagnosis and Treatment. *Rheumatology and Therapy*, **3**, 187-207. <https://doi.org/10.1007/s40744-016-0040-4>
- [2] Still, G.F. (1897) On a Form of Chronic Joint Disease in Children. *Medico-Chirurgical Transactions*, **80**, 47. <https://doi.org/10.1177/095952879708000106>
- [3] Maldonado-Cocco, J.A., García-Morteo, O., Spindler, A.J., Hübscher, O. and Gagliardi, S. (1980) Carpal Ankylosis in Juvenile Rheumatoid Arthritis. *Arthritis & Rheumatism: Official Journal of the American College of Rheumatology*, **23**, 1251-1255. <https://doi.org/10.1002/art.1780231104>
- [4] Petty, R.E., Southwood, T.R., Manners, P., *et al.*, (2004) International League of Associations for Rheumatology Classification of Juvenile Idiopathic Arthritis: Second Revision, Edmonton, 2001. *The Journal of rheumatology*, **31**, 390.
- [5] Weiss, J.E. and Ilowite, N.T. (2007) Juvenile Idiopathic Arthritis. *Rheumatic Disease Clinics of North America*, **33**, 441-470. <https://doi.org/10.1016/j.rdc.2007.07.006>
- [6] Gowdie, P.J. and Tse, S. (2012) Juvenile Idiopathic Arthritis. *Pediatric Clinics of North America*, **59**, 301-327. <https://doi.org/10.1016/j.pcl.2012.03.014>
- [7] Weiss, J.E. and Ilowite, N.T. (2005) Juvenile Idiopathic Arthritis. *Pediatric Clinics of North America*, **52**, 413-442. <https://doi.org/10.1016/j.pcl.2005.01.007>
- [8] Prakken, B., Albani, S. and Martini, A. (2011) Juvenile Idiopathic Arthritis. *The Lancet*, **377**, 2138-2149. [https://doi.org/10.1016/S0140-6736\(11\)60244-4](https://doi.org/10.1016/S0140-6736(11)60244-4)
- [9] Peterson, L.S., Mason, T., Nelson, A.M., O'fallon, W.M. and Gabriel, S.E. (1996) Juvenile Rheumatoid Arthritis in Rochester, Minnesota 1960-1993. Is the Epidemiology Changing? *Arthritis & Rheumatism*, **39**, 1385-1390. <https://doi.org/10.1002/art.1780390817>
- [10] Eisenstein, E.M. and Berkun, Y. (2014) Diagnosis and Classification of Juvenile Idiopathic Arthritis. *Journal of Autoimmunity*, **48**, 31-33. <https://doi.org/10.1016/j.jaut.2014.01.009>
- [11] Manners, P.J. and Diepeveen, D.A. (1996) Prevalence of Juvenile Chronic Arthritis in a Population of 12-Year-Old Children in Urban Australia. *Pediatrics*, **98**, 84-90. <https://doi.org/10.1097/01241398-199701000-00060>
- [12] Saurenmann, R., Rose, J., Tyrrell, P., *et al.* (2007) Epidemiology of Juvenile Idiopathic Arthritis in a Multiethnic Cohort: Ethnicity as a Risk Factor. *Arthritis & Rheumatism*, **56**, 1974-1984. <https://doi.org/10.1002/art.22709>
- [13] Aggarwal, A. and Misra, R. (1996) Juvenile Rheumatoid Arthritis in India—Rarity of Antinuclear Antibody and Uveitis. *The Indian Journal of Pediatrics*, **63**, 301-304. <https://doi.org/10.1007/BF02751522>
- [14] El-Soud, A.M.A., El-Najjar, A.R., El-Shahawy, E.E., *et al.* (2013) Prevalence of Juve-

- nile Idiopathic Arthritis in Sharkia Governorate, Egypt: Epidemiological Study. *Rheumatology International*, **33**, 2315-2322. <https://doi.org/10.1007/s00296-013-2707-2>
- [15] Abdwani, R., Abdalla, E., Al Arawi, S. and Al-Zakwani, I. (2015) Epidemiology of Juvenile Idiopathic Arthritis in Oman. *Pediatric Rheumatology*, **13**, 33. <https://doi.org/10.1186/s12969-015-0030-z>
- [16] Ravelli, A., Felici, E., Magni-Manzoni, S., *et al.*, (2005) Patients with Antinuclear Antibody-Positive Juvenile Idiopathic Arthritis Constitute a Homogeneous Subgroup Irrespective of the Course of Joint Disease. *Arthritis & Rheumatism*, **52**, 826-832. <https://doi.org/10.1002/art.20945>
- [17] Woerner, A., von Scheven-Gête, A., Cimaz, R. and Hofer, M. (2015) Complications of Systemic Juvenile Idiopathic Arthritis: Risk Factors and Management Recommendations. *Expert Review of Clinical Immunology*, **11**, 575-588. <https://doi.org/10.1586/1744666X.2015.1032257>
- [18] Yfantopoulos, J. and Sarris, M. (2001) Health Related Quality of Life. *Measurement Methodology. Archives of Hellenic Medicine*, **18**, 218-229.
- [19] Luca, N.J. and Feldman, B.M. (2014) Health Outcomes of Pediatric Rheumatic Diseases. *Best Practice & Research Clinical Rheumatology*, **28**, 331-350. <https://doi.org/10.1016/j.berh.2014.04.001>
- [20] Calvert, M., Blazeby, J., Altman, D.G., *et al.* (2013) Reporting of Patient-Reported Outcomes in Randomized Trials: The Consort PRO Extension. *JAMA*, **309**, 814-822. <https://doi.org/10.1001/jama.2013.879>
- [21] Madi, S.M., Al-Mayouf, S.M., Grainger, C.G. and Bahabri, S.A. (2004) The Arabic Version of Childhood Health Assessment Questionnaire Modified for Arabic Children. *Saudi Medical Journal*, **25**, 83-87.
- [22] Hays, R.D., Staquet, M.J. and Fayers, P.M. (1998) Quality of Life Assessment in Clinical Trials: Methods and Practice. Oxford University Press, Oxford.
- [23] Amine, B., Rostom, S., Benbouazza, K., Abouqal, R. and Hajjaj-Hassouni, N. (2009) Health Related Quality of Life Survey about Children and Adolescents with Juvenile Idiopathic Arthritis. *Rheumatology International*, **29**, 275-279. <https://doi.org/10.1007/s00296-008-0672-y>
- [24] Shaaban, F.A., Metwally, I.M., Samy, S.M., Salama, I.I. and Hassanin, A.I. (2006) Health Related Quality of Life, Disease Activity, Severity and Coping in Juvenile Rheumatoid Arthritis. *Journal of Medical Sciences*, **6**, 561-568. <https://doi.org/10.3923/jms.2006.561.568>
- [25] Mańczak, M., Rutkowska-Sak, L. and Raciborski, F. (2016) Health-Related Quality of Life in Children with Juvenile Idiopathic Arthritis—Child's and Parent's Point of View. *Reumatologia*, **54**, 243-250. <https://doi.org/10.5114/reum.2016.63665>
- [26] Oliveira, S., Ravelli, A., Pistorio, A., *et al.* (2007) Proxy-Reported Health-Related Quality of Life of Patients with Juvenile Idiopathic Arthritis: The Pediatric Rheumatology International Trials Organization Multinational Quality of Life Cohort Study. *Arthritis Care & Research*, **57**, 35-43. <https://doi.org/10.1002/art.22473>
- [27] Ravelli, A. (2004) Toward an Understanding of the Long-Term Outcome of Juvenile Idiopathic Arthritis. *Clinical and Experimental Rheumatology*, **22**, 271-275.
- [28] Laaksonen, A.-L. (1966) A Prognostic Study of Juvenile Rheumatoid Arthritis. Analysis of 544 Cases. *Acta Paediatrica Scandinavica*, **166**, 49-55.
- [29] Meiorin, S., Filocamo, G., Pistorio, A., *et al.* (2009) Impact of Involvement of Individual Joint Groups on Subdimensions of Functional Ability Scales in Juvenile

Idiopathic Arthritis. *Clinical & Experimental Rheumatology*, **27**, 527.

- [30] Ravelli, A. and Martini, A. (2007) Juvenile Idiopathic Arthritis. *The Lancet*, **369**, 767-778. [https://doi.org/10.1016/S0140-6736\(07\)60363-8](https://doi.org/10.1016/S0140-6736(07)60363-8)
- [31] Sawyer, M.G., Whitham, J.N., Roberton, D.M., *et al.* (2004) The Relationship between Health-Related Quality of Life, Pain and Coping Strategies in Juvenile Idiopathic Arthritis. *Rheumatology (Oxford)*, **43**, 325-330. <https://doi.org/10.1093/rheumatology/keh030>
- [32] Cassidy, J.T., Levinson, J., Bass, J., *et al.*, (1986) A Study of Classification Criteria for a Diagnosis of Juvenile Rheumatoid Arthritis. *Arthritis & Rheumatism: Official Journal of the American College of Rheumatology*, **29**, 274-281. <https://doi.org/10.1002/art.1780290216>
- [33] Gotte, A.C. (2009) Intra-Articular Corticosteroids in the Treatment of Juvenile Idiopathic Arthritis: Safety, Efficacy, and Features Affecting Outcome. A Comprehensive Review of the Literature. *Open Access Rheumatology: Research and Reviews*, **1**, 37. <https://doi.org/10.2147/OARRR.S5103>
- [34] Beukelman, T., Patkar, N.M., Saag, K.G., *et al.* (2011) 2011 American College of Rheumatology Recommendations for the Treatment of Juvenile Idiopathic Arthritis: Initiation and Safety Monitoring of Therapeutic Agents for the Treatment of Arthritis and Systemic Features. *Arthritis Care & Research*, **63**, 465-482. <https://doi.org/10.1002/acr.20460>
- [35] Céspedes-Cruz, A., Gutiérrez-Suárez, R., Pistorio, A., *et al.* (2008) Methotrexate Improves the Health-Related Quality of Life of Children with Juvenile Idiopathic Arthritis. *Annals of the Rheumatic Diseases*, **67**, 309-314. <https://doi.org/10.1136/ard.2007.075895>
- [36] Hertzberger-ten, R.C. and Cats, A. (1991) Toxicity of Sulfasalazine in Systemic Juvenile Chronic Arthritis. *Clinical and Experimental Rheumatology*, **9**, 85-88.
- [37] Stoll, M.L. and Cron, R.Q. (2014) Treatment of Juvenile Idiopathic Arthritis: A Revolution in Care. *Pediatric Rheumatology*, **12**, 13. <https://doi.org/10.1186/1546-0096-12-13>
- [38] Robinson, R.F., Nakata, M.C., Hayes, J.R., Rennebohm, R. and Higgins, G. (2003) Quality-of-Life Measurements in Juvenile Rheumatoid Arthritis Patients Treated with Etanercept. *Clinical Drug Investigation*, **23**, 511-518. <https://doi.org/10.2165/00044011-200323080-00003>