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ORIGINAL ARTICLE

Quality of life in children and adults with Idiopathic juvenile arthritis: cross-sectional study in Brazilian patients

Qualidade de vida em crianças e adultos com artrite idiopática juvenil: estudo transversal em pacientes brasileiros

Elora Sampaio Lourenço[1], Cibele Lourenço[1], Thelma Skare[1], Paulo Spelling[1], Renato Nisihara[1]

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ABSTRACT

BACKGROUND AND OBJECTIVES: Juvenile idiopathic arthritis (JIA) is a chronic rheumatic disease which may persist into adulthood. Pain and joint deformities affect quality of life (QoL). The objective was to study the influence of JIA in QoL of children and adults, comparing the two groups.

METHODS: Cross-sectional study of 47 JIA patients (20 children and 27 adults) using the following questionnaires: Juvenile Arthritis Multidimensional Assessment Report (JAMAR), Health Assessment Questionnaire (HAQ) in adults and Childhood Health Assessment Questionnaire CHAQ) in children. Epidemiological, clinical and treatment data was collected.

RESULTS: No differences were found in gender, arthritis subset, presence of uveitis and fever, visual analog scale for pain and duration of morning stiffness. Adult patients required more biologics in the treatment (p=0.02) and had higher age at disease onset than children (p=0.001). About 45% of children and 51.8% of adults complained having pain and 40% of children and 48% of adults were not satisfied with the current status of their disease.

CONCLUSION: Children and adult patients with JIA had similar degree of pain, morning stiffness and functional impairment. Functional impairment was considered mild but the level of pain found was higher than in other studies. Almost half of patients were not satisfied with the treatment.

Keywords: Adult, Child, Juvenile arthritis, Pain.

RESUMO

JUSTIFICATIVA E OBJETIVOS: A artrite idiopática juvenil (AIJ) é uma doença reumática crônica que pode persistir na idade adulta. Dor e deformidades articulares afetam a qualidade de vida (QV). O objetivo foi estudar a influência da AIJ na QV de crianças e adultos, comparando os dois grupos.

MÉTODOS: Estudo transversal de 47 pacientes com AIJ (20 crianças e 27 adultos) utilizando os questionários: *Juvenile Arthritis Multidimensional Assessment Report* (JAMAR), *Health Assessment Questionnaire* (HAQ) em adultos e *Childhood Health Assessment Questionnaire* – (CHAQ) em crianças. Dados epidemiológicos, clínicos e de tratamento foram coletados.

RESULTADOS: Não foram encontradas diferenças em sexo, subconjunto de artrite, presença de uveíte e febre, escala analógica visual para dor e duração da rigidez matinal. Pacientes adultos necessitaram mais fármacos biológicos no tratamento (p=0,02) e tinham maior idade de início da doença do que crianças (p=0,001). Cerca de 45% das crianças e 51,8% dos adultos queixaram-se de dor e 40% das crianças e 48% dos adultos não estavam satisfeitos com o estado atual de sua doença.

CONCLUSÃO: Pacientes crianças e adultos com AIJ apresentaram graus semelhantes de dor, rigidez matinal e comprometimento funcional. O comprometimento funcional foi considerado leve, mas o nível de dor encontrado foi maior do que em outros estudos. Quase metade dos pacientes não ficou satisfeita com o tratamento.

Descritores: Adulto, Artrite juvenil, Criança, Dor.

INTRODUCTION

The most common rheumatic disease in childhood is juvenile idiopathic arthritis (JIA), which has disease onset before 16 years of age and unknown etiology[1]. JIA is an umbrella term that encompasses several entities that have diverse pathophysiologic processes, clinical features and prognostic outcomes. Among them are the polyarticular forms positive for rheumatoid factor (RF), polyarticular form with negative RF, oligoarticular forms, psoriatic, systemic, enthesitis-related arthritis and a seventh category, the undifferentiated arthritis, which includes those patients who do not fit any criteria or fit more than one[2].

Patients with JIA may have lower quality of life (QoL) than normal population[3]. Chronic pain and physical restrictions limiting participation at school, sports activities and social events are some of the contributors to the reduction of well-being. Loss of

Elora Sampaio Lourenço – https://orcid.org/0000-0002-3281-0433;
Cibele Lourenço – https://orcid.org/0000-0003-1602-6304;
Thelma Skare – https://orcid.org/0000-0002-7699-3542;
Paulo Spelling – https://orcid.org/0000-0002-1980-4645;
Renato Nisihara – https://orcid.org/0000-0002-0297-6731.

1. Mackenzie Evangelical School of Medicine, Medicine, Curitiba, PR, Brazil.

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Correspondence to:
Renato Nisihara
Faculdade Evangélica Mackenzie do Paraná.
Rua Padre Anchieta, 2770
80730-000 Curitiba, PR, Brasil.
email: renatonisihara@gmail.com; renato.nisihara@fempar.edu.br

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QoL may occur even when the inflammatory activity is well controlled[3] due to functional disabilities secondary to joint damage, deformities and drugs' side effects such as low bone mass and growth restriction[4]. Growth disturbances and joint contractures may also impact in the corporal image bringing psychological problems and social isolation. Extra articular manifestations, mainly uveitis, which can be sight threatening and affects up to 10% of this population, may further aggravate the situation[1]. Therefore, the ideal treatment in JIA should include a multidisciplinary team.

Some children, mainly those with oligoarticular JIA, accomplish remission[1], but almost half of JIA patients may have the active disease during adulthood[4] and many will have some restriction in their activities of daily living[5].

The therapeutic approach to JIA has changed with the introduction of new drugs and strategies that seek to obtain the best control of inflammatory activity[1,5]. Nevertheless, the study[6] showed that 76% of the children reported having pain in at least 60% of the days and that pain perception is closely related to functional status and mood and sleep alterations, reducing QoL.

The present study evaluated children and adult patients with JIA from Brazil aiming to know the influence of this disease in their QoL and establishing a comparison between these two groups. The hypothesis is that adult patients with JIA, having a disease of longer duration and that remained active during adulthood would have more pain and worst QoL than children with JIA.

METHODS

A cross-sectional study including patients older than 6 years of age that fulfilled the classification criteria for JIA from International League of Associations for Rheumatology (ILAR)[7]. Patients with cognitive deficiencies and with associated chronic diseases that could interfere in QoL and pain were excluded.

This was a convenience sample that included all patients seen in the Pediatric Rheumatology Unit and Adult Outpatient Rheumatology Clinics from the same hospital that filled the inclusion criteria and agreed to participate in the study.

A total of 47 patients were included: 20 from the Pediatric Rheumatologic Unit and 27 from the adult Rheumatologic Unit. Epidemiological and clinical data were obtained through chart reviews. All patients were submitted to Juvenile Arthritis Multidimensional Assessment Report (JAMAR)[8]. The Childhood Health Assessment Questionnaire (CHAQ) was applied to children[9] and adults answered the Health Assessment Questionnaire (HAQ)[10]. All applied instruments were translated and validated to Portuguese language[8-10].

JAMAR[8] is a questionnaire that allows multidimensional evaluation of JIA. It's auto applicable in individuals over 7 years of age and has 15 domains that evaluate the following: functional status measured through the Juvenile Arthritis Functionality Scale (JAFS), which has several questions on daily life activities and is graded from 0 (no difficulties) to 45 (disability); pain intensity- using a visual analog scale (VAS) whose score ranges from 0 (no pain) to 10 (worst scenario); patients' perception of articular pain and edema; degree of morning stiffness; extra articular symptoms; patients' perception of disease activity graduated from 0 (no activity) to 10 (maximum activity); evaluation of disease course at the moment of interview: remission, persistent activity or relapse; classification of the disease status in relation to previous consultation (from much better to much worse); drug treatment; treatment side effects; treatment adhesion; degree of difficulties in work/school caused by the disease; evaluation of QoL through the Pediatric Rheumatology Quality of Life Scale (PRQL), which is a 10-item measurement of QoL that uses a 4-point Likert scale ranging from 0 (never) to 3 (all the time) and a total score from 0 to 30, with higher scores indicating worse quality of life; evaluation of general well-being through the VAS (from 0 =very well to 10=very bad); a question on the degree of satisfaction with the present status of the disease (with yes or no response).

The HAQ[10] is an instrument used to evaluate functional status in adults that has 20 questions on activities of daily living and ranges from zero (no impairment) to 3 (worst scenario); the CHAQ[9] is a HAQ version modified for children with 30 items that also ranges from zero (no impairment) to 3 (worst scenario). Results under one are considered to be secondary to a light impairment and values over two to severe impairment[11].

The obtained data was analyzed using frequency and contingency data. Exact Fisher and Chi-squared tests were used to compare nominal and categorical data (gender, clinical disease subsets, presence of uveitis, antinuclear antibody and rheumatoid factor, used treatment and their side effects, presence of fever, skin rash and pain in the last week, morning stiffness category, categories of patient's perception in the disease evolution and in difficulties found in daily life).

This study was approved by the local Committee of Ethics in Research under protocol number 2.901.915; all patients and/or their guardians signed the Free and Informed Consent Term (FICT).

Statistical analysis

Mann-Whitney and unpaired t tests were used to compare numerical data age at disease onset, values of JAFS, PRQL and VAS of pain and well-being and VAS of perception in the disease activity. The adopted significance was of 5%. Calculus was done with the help of the software Graph Pad Prism version 6.00 for Windows, GraphPad Software, La Jolla, California, USA, www.graphpad.com.

RESULTS

The sample included 47 individuals: 20 children (mean age of 11.7±2.8 years) and 27 adults (mean age of 28.1±9.4 years). Females comprised 75% of children's and 70.4% of adults' subgroup.

This sample had preponderance of polyarticular forms of JIA (60% of children vs 70.3% of adults) and the studied children and adults had the same proportion of the disease subsets (p=0.45), allowing a valid comparison. Patients that persisted with JIA into adulthood had later disease onset (children at 5.85±3.9 years and adults at 10.0±4.4 years with p=0.001) and used more biologics as treatment (30% vs 66.6%; p=0.02), while there were more children without drug treatment (40% vs 7.4%). No differences in use of non-steroidal anti-inflammatory drugs (NSAIDs), other disease modifying drugs and glucocorticoid were note (all with p>0.05). Also, no differences in VAS of pain in the last week. Fever and skin rashes were found (all with p>0.05) but adults had more morning stiffness than children (p=0.02).

The comparison of the degree of impairment measured in children by CHAQ and adults by HAQ is presented in table 1. This table shows that most of individuals in the two groups had mild impairment with no differences between children and adults.

Table 1. Comparison of functional indexes in juvenile idiopathic arthritis in children and adults

Impairment	Children n=20	Adults n=27	p-value
Mild	18 (90%)	18 (66.6%)	0.17
Moderate	1(5%)	5 (18.5%)	
Severe	1 (5%)	4 (14.8%)	

The comparison of functionality assessed by the JAFS component of JAMAR and QoL measured by the PRQL component of JAMAR were observed and the results on table 2 were found.

Table 2. Functionality, quality of life and patients' perception on disease present status and evolution in 47 patients with juvenile idiopathic arthritis.

	Children n=20	Adults n=27	p-value
JAFS	Range: 0 - 2.8 Median 0.6 (0.2 - 1.7)	Range: 0 - 4.8 Median 1.5 (0.6 - 3.1)	0.10
PRQL	Range: 0 - 5.6 Median 1.3 (0.7 - 1.9)	Range: 0 - 8 Median 2.3 (0.6 - 4.6)	0.13
Median of patients' perception of intensity of disease activity	4.0 (0 - 5.0)	4.0 (0 - 7.0)	0.86
Median of intensity of patients' well-being	2.5 (0 - 5.0)	4.0 (0 - 6.0)	0.63
Patients' perception evolution of disease			
Remission	6 (30.0%)	6 (22.2%)	0.80
Persistent activity	6 (30.0%)	10 (37.0%)	
Relapse	8 (40.0%)	11 (40.7%)	
Patients' comparison of the disease in relation to previous consultation			
Good improvement	4 (20.0%)	4 (14.8%)	0.16
Light improvement	7 (35.0%)	3 (11.1%)	
Stable	8 (40.0%)	16 (59.2%)	
Little worsening	1 (5.0%)	4 (14.8%)	
Important worsening	-	-	

n = number; JAFS = Juvenile Arthritis Functionality Scale; PRQL = Pediatric Rheumatology Quality of Life Scale; Between brackets-interquartile range.

In this table it's possible to see that the comparison of patients' perception in the disease evolution was the same. Also, no differences were found in functionality, QoL, well-being and patient's perception in the disease activity.

Table 3 shows the main problems found at school and work and the degree of satisfaction with the present status of the disease. Absenteeism and difficulties to remain seated were the most common.

Table 3. Difficulties found at school/work and degree of satisfaction with the present status of the disease in 47 juvenile idiopathic arthritis patients

	Children n=20	Adults* n=17	p-value
No difficulties	8 (40.0%)	8 (47.0%)	0.75
Absenteeism	9 (45.0%)	7 (41.1%)	0.91
Difficulties to remain seated	6 (30.0%)	4 (23.5%)	0.72
Difficulties related to teachers/colleagues	1 (5.0%)	1 (5.8%)	1.0
Decreasing production at work/school	3 (15.0%)	5 (29.4%)	0.42
Satisfaction with present status (Yes/No)	12/8	14/13	0.57

* 10 adults reported they were not studying or working at the time of the interview.

DISCUSSION

Few Brazilian papers addressed the quality of life in JIA patients. In the present study, it was found that adult Brazilian patients with JIA had later disease onset and required more biologic treatment than children. It was not possible to find a study that established a similar comparison of age of onset in this context. This is a finding that could help predict those with persistence of disease into adulthood. However, more studies with higher number of patients are needed to replicate this finding.

Very few differences in the degrees of pain, stiffness and functionality were found when the group of adults and children were compared. Study[12], done in 1997, showed that AIJ patients with longer disease duration had greater functional disability and pain so it could be expected that adult patients had worse scores than children. A review[13] of 984 JIA patients with mean age of 30 years showed that 47% of patients still had active arthritis and 46% complained of difficulties in daily living.

In the present study, the degree of functional impairment found could be considered low, with mean values of 0.6 for children and 1.5 for adults in the JASF instrument, which is consistent with the findings of HAQ and CHAQ, where the majority of individuals had mild impairment. It's possible that the use of biologics and an aggressive treatment to avoid inflammation adopted nowadays have changed this aspect. This treatment approach may also have blunted the observation of other differences such as in pain and stiffness between the two groups.

Nevertheless, about 40% (8/20) of children and 48% (13/27) of adults were not satisfied with the current status of the disease, showing that there is a need for further improvement in the treatment. One item that should be addressed in this context is the pain treatment that was found to have a VAS median value of 4 for adults and 2.5 for children. These results are higher than those found in the works[14,15], that found median values of 2.5 and zero, respectively, while studying children. They were also higher than those of the study[16], that evaluated only adults with JIA and found a median VAS of 2.3. Therefore, the present findings suggest that there is a need for improvement in pain treatment. Corroborating this idea, almost half of adults and children reported having had pain in the last week.

Pain in JIA is poorly understood as the inflammatory activity, although influential, it accounts for just a small proportion in variance of pain in these patients[17]. It has been shown that children with arthritis continue to experience clinically significant pain despite adequate doses of disease-modifying antirheumatic drugs and anti-inflammatory agents[18]. In this context, pain may be related with amplified sensitivity to painful mechanical and thermal stimuli, even in absence of markers of disease activity[19]. Unluckily, most of the current studies in this issue are focused on examining relationships between pain and other variables rather than examining its nature[17]. Older age at disease onset, poor function/disability and longer disease duration at baseline were associated consistently with high degree of pain. It's believed that older children may have difficulties to adjust with the changes to their functioning than the younger ones and they may also perceive the long-term consequences of JIA as more important[17].

When social difficulties were analyzed, problems related to school and work were the most common, mainly absenteeism, which needs to be addressed by the attending physician in order to be minimized. Going to school helps developing not only cognitive but also social skills and the degree of school absenteeism is associated with isolation, anxiety and lower educational level[20]. In adults, a study[5] has shown that, although these patients have the same educational achievement than controls, the rate of unemployment in this group was 3 times higher than controls.

This study also showed that adult patients received more treatment with biologics. The higher requirement of biologics in adults may reflect the higher severity of the persistent disease. It's also possible that this circumstance was only due to longer disease duration in adults. However, different approaches from adult and pediatric rheumatologists may have had some influence. There is no guidance for treatment of adult patients with JIA and most of adult rheumatologists treat them as rheumatoid arthritis (RA) that may not be appropriated, as JIA is a more heterogeneous disease. In JIA, treatment should follow the disease subtype. Unluckily, most of the drug trials group different subtypes together in order to obtain a large sample size, missing important differences among them. Methotrexate is usually the first drug used as disease modifying for peripheral arthritis, and the most frequently prescribed biologic are the anti TNF-alpha drugs that have been shown to be effective and safe in the majority of cases. However, more recently, the choice of biologics has extended to include alternative cytokine blockage and abatacept. These last options may be effective in polyarticular and systemic forms of the disease but are not adequate for the others[21].

The present study has several limitations: the small number of the sample and its cross-sectional design are some of them. In addition, the sample had only oligoarticular and polyarticular forms of the disease, not representing its complete spectrum. Also, it studied patients treated in a tertiary hospital that may not represent the general population, which may have higher complications rate of the disease due to difficulties in the treatment access. Moreover, despite improvement in functional outcome achieved in the last years, a great proportion of JIA patients are still unsatisfied with treatment. An attempt at a better treatment of pain is one of the strategies that could improve their degree of satisfaction.

CONCLUSION

Children and adult patients with JIA had similar degree of pain, morning stiffness and functional impairment. Functional impairment was considered mild but the level of pain found was higher than in other studies. Almost half of patients were not satisfied with the treatment.

AUTHORS' CONTRIBUTIONS

Elora Sampaio Lourenço
Data Collection, Conceptualization, Research, Writing - Preparation of the original, Writing - Review and Editing
Cibele Lourenço
Data Collection, Conceptualization, Methodology, Writing - Preparation of the original, Writing - Review and Editing
Thelma Skare
Statistical analysis, Data collection, Conceptualization, Research, Methodology, Writing - Preparation of the original, Writing - Review and Editing
Paulo Spelling
Data Collection, Conceptualization, Writing - Preparation of the original, Writing - Review and Editing
Renato Nisihara
Conceptualization, Research, Methodology, Writing - Preparation of the original, Writing - Review and Editing, Supervision

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