ECRONICON OPEN ACCESS

EC PAEDIATRICS
Research Article

Assessing the Clinical Pattern of JIA Presented in a Tertiary level Hospital

Mamun Miah[1]*, Md Tarek Azad[2], Kazi Zahidul Hoque[3], Akhand Tanzih Sultana[4], Shubhra Prakash Paul[5] and Md Zahangir Alam[6]

[1]Associate Professor, Department of Paediatric Rheumatology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh

[2]Professor and Head Department of paediatrics, Jalalabad Ragib Rabeya Medical College, Sylhet, Bangladesh

[3]Assistant Professor and Unit Chief, Department of Paediatric Cardiac Surgery, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh

[4]Assistant Professor, Department of Respiratory Medicine, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh

[5]Lecturer, Community Medicine, Rajshahi Medical College, Rajshahi, Bangladesh

[6]Professor and Head Department of Paediatric Rheumatology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh

***Corresponding Author**: Mamun Miah, Associate Professor, Department of Paediatric Rheumatology, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka, Bangladesh.

Received: November 04, 2019; **Published:** November 19, 2019

Abstract

Background and Objective: Juvenile Idiopathic Arthritis (JIA) is a disease that shows wide variation in different populations. The International League of Associations for Rheumatology (ILAR) classification has been used all over the world for delineate JIA disease characteristics in various population. As these variations may provide insight into the cause of JIA, there is substantial interest in the differences in JIA to provide JIA subtype specific management plan to control the disease in the community. The present study was, therefore, undertaken to see the JIA profile in Bangladeshi children.

Materials and Methods: This descriptive study was conducted in Dhaka Shishu (Children) Hospital and Bangladesh Institute of Child Health over a period of 2 and a half year between July 2014 to December 2016. Prior to study, ethical clearance was obtained from Ethical Review Committee of the Institute. A total of 52 children diagnosed as JIA as per the ILAR 2001 criteria were the study sample. Data were collected using a semi-structured questionnaire containing the variables of interest which among others included demographics, disease subtype, and routine blood results. A doubly-anchored horizontal 100 mm visual analogue scale (VAS) was used for the assessment of the child's overall wellbeing and a doubly-anchored horizontal 100 mm VAS for the assessment of the intensity of the child's pain.

Results: Majority (80.8%) of the children was diagnosed as having JIA at the age of 5 - 10 years. The mean ages at onset and presentation of the disease were 7.5 and 7.9 years respectively. A male predominance was observed in the series. The predominant JIA subtype was rheumatoid factor (RF) negative polyarthritis (42.3%) followed by oligo-persistent (25%) and RF positive polyarthritis (11.5%). Oligo-extended and systemic onset JIA each comprised of 7.7%. Enthesitis-related arthritis (ERA) was rare (5.8%). Disease activities of the children at presentation showed that the mean intensity of pain on VAS was 35.8 (range: 25 - 50) mm. Children's overall well-being on VAS, was on an average 25.2 (range: 20 - 40) mm. The mean numbers of active joints and number of limited joints were 4.0 (range: 2 - 8) and 2.0 (range: 0 - 4) respectively.

Conclusion: The present study revealed a different JIA profile compared to other international JIA studies. JIA was predominant in male children. RF negative polyarthritis is more common followed by oligo-persistent. ERA was seldom observed.

Keywords: *Juvenile Idiopathic Arthritis; Subtype; Clinical Features*

Introduction

Juvenile Idiopathic Arthritis (JIA) is the most common chronic arthritis in children worldwide. According to the International League of Associations for Rheumatology (ILAR) [1], JIA is defined as arthritis that begins before the 16th birth anniversary and persists for at least 6 weeks, provided other conditions being excluded [2]. It is not a single disease entity but a heterogeneous group of chronic arthritic disorders of unknown cause in children. JIA is an important cause of short- and long-term disability in children with decreased daily function and quality of life [3,4].

The ILAR classification has, in recent years, been used all over the world to delineate JIA disease characteristics in various populations and nationalities. Included in these recent studies are large multicentre trials and smaller studies from both developed and developing countries [5-9]. Studies in developed countries have estimated a prevalence of JIA that varies between 0.07 - 4 per 1000 children [10,11]. Reports on the incidence and prevalence of JIA suggest variability among different ethnic and geographically distinct populations [12,13]. As evidence increasingly shows, it is likely that JIA prevalence is underestimated.

In Bangladesh, rural people and urban slum dwellers have poor access to tertiary level or specialized healthcare, and as such, accurate magnitude of the disease is not available. The first large community-based study (carried out among 16,270 children who were selected by multistage sampling technique from a community of approximately 1,05,986 children in the Narayanganj district, Bangladesh between November 2008 to December 2009) published in 2012 showed that the prevalence of JIA was 60.5 per 100000 children in rural area with girl to boy ratio being 2.3:1.0. The subgroup distribution showed oligoarticular JIA in the majority of patients (60%) [14]. A hospital-based study conducted in Bangabandhu Sheikh Mujib Medical University (BSMMU) between July 2007 - December 2012 by Islam., *et al*. demonstrated that a total of 540 patients of arthritis were enrolled during the study period. Of them JIA was predominant (77%) followed by SLE (10%), vasculitis (6.7%) (which comprised of Henoch Shonlein purpura (HSP), polyarteritis nodosa, Kawasaki Disease and other rare varieties of juvenile arthritis) [15]. As these variations may provide insight into the cause of JIA, there is substantial interest in the differences in JIA in various parts of the world. The present study is conducted to find the pattern of JIA in the urban setting of a tertiary care hospital.

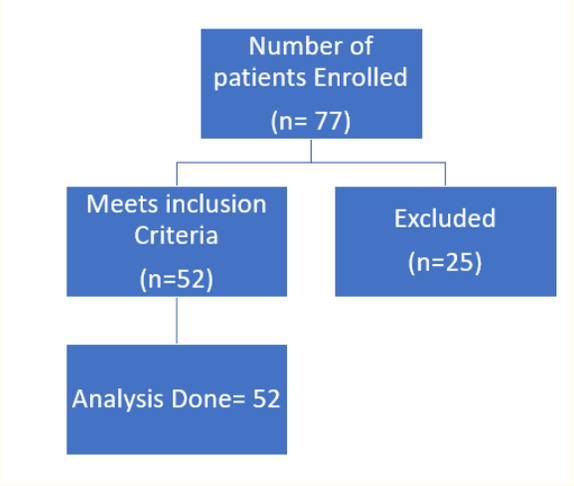
Materials and Methods

This descriptive study was conducted in Dhaka Shishu Hospital and Bangladesh Institute of Child Health over a period of 2 and a half years between July 2014 to December 2016. Prior to study Ethical clearance was obtained from Ethical Review Committee of the Institute. The children diagnosed as JIA as per the ILAR 2001 criteria [1] were the study population. Only those patients whose parents gave written consent to participate in the study were enrolled and those patients are selected for the study who are suffering from JIA, and excluded patients were those who have other comorbidities along with JIA. Data were collected using a semi-structured questionnaire containing the variables of interest which among others included demographics, disease subtype, and routine blood results. Blood tests were done only if necessary, for routine medical care and previous relevant blood results were collected from the case notes. It was not routine to perform blood tests for Anti-nuclear antibody (ANA), Human leukocyte antigen B27 (HLAB27) at every visit due to resource limitations and unavailability. ANA test was only done (using ELISA method) in children who presented with oligoarthritis, HLAB27 was only done in those with other features of enteritis-related arthritis (ERA) and RF was done only in children with polyarthritis but this was not part of our study, therefore, it was not included in this article.

A doubly-anchored horizontal 100 mm visual analogue scale (VAS) was used for the assessment of the child's overall wellbeing (with anchors of '0 = very well' and '100 = very poor') and a doubly-anchored horizontal 100 mm VAS for the assessment of the intensity of the child's pain (with anchors of '0 = no pain' and '100 = very severe pain') [5]. The CHAQ is a well validated international tool and has been used in many recent JIA trials [16]. Data were analyzed using SPSS (Statistical Package for Social Sciences), version 17 and test statistics used to analyse the data were descriptive statistics. While the categorical data were presented as absolute number and percentage, the continuous data were presented as mean/median and standard deviation (SD) from the mean.

Results

Total number of patients admitted during the study period = 200



Figure

Majority (80.8%) of the children was diagnosed as having JIA at the age of 5 - 10 years and only a few cases below the age 5 years. The mean ages at onset and presentation of the disease were 7.5 and 7.9 years respectively. Males outnumbered females roughly by 3:2. Over half (53.8%) of the children at presentation had been suffering from the disease for 6 or < 6 months and 42.3% for 7 - 12 months. Only 2 (3.8%) children had disease duration of > 12 months (Table 1). Figure 1 shows the subtypes of JIA in our series. The predominant JIA subtype was rheumatoid factor (RF) negative polyarthritis (42.3%) followed by oligo-persistent (25%) and RF positive polyarthritis (11.5%). Oligo-extended and systemic onset JIA each comprised of 7.7% and enthesitis-related arthritis (ERA) 5.8% of the cases. Disease activities of the children at presentation are illustrated in table 2. The mean intensity of pain measured in visual analogue scale (0 - 100 mm) was 35.8 (range: 25 - 50) mm. Children's overall well-being, also measured in VAS (0 - 100 mm), was on an average 25.2 (range: 20 - 40) mm. The mean number of active joints and number of joints with restricted movement were 4.0 (range: 2 - 8) and 2.0 (range: 0 - 4) respectively.

Demographic characteristics	Frequency	Percentage	Mean ± SD (range)
Age at presentation (years)			
< 5	02	3.8	7.9 ± 2.8 (4 - 15)
5 - 10	42	80.8	
> 10	08	15.4	
Age at onset (years)	---	---	7.5 ± 2.6 (4 - 15)
Sex			
Male	30	57.7	---
Female	22	42.3	---
Duration of illness (months)			
≤ 6	28	53.8	7.6 ± 4.4 (2 - 23)
7 - 12	22	42.3	
> 12	02	3.8	

Table 1: *Distribution of children by their demographic characteristics (n = 52).*

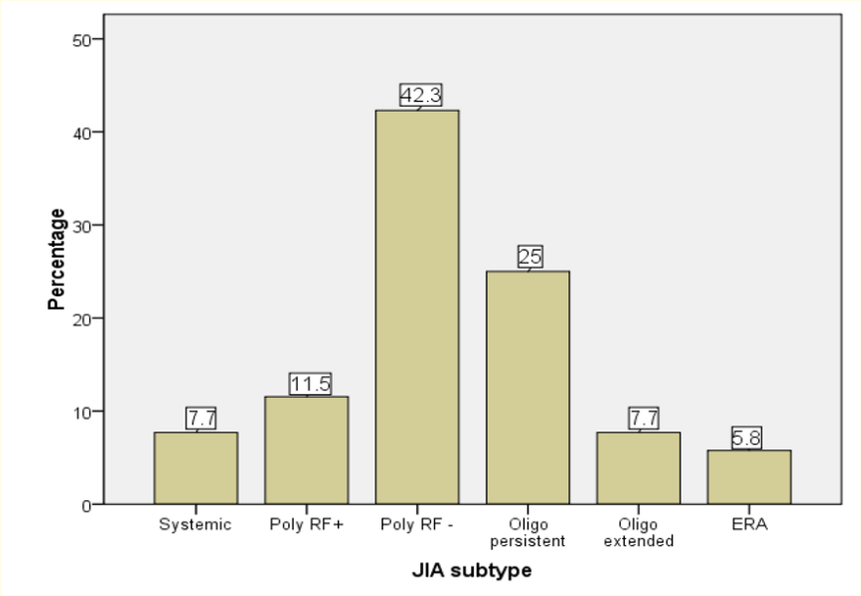


Figure 1: *The subtypes of JIA in our series.*

1. Systemic: Systemic onset JIA, 2. Poly RF: Polyarticular JIA Rheumatoid Factor Positive, 3. Poly RF: Polyarticular JIA Rheumatoid Factor Negative, 4. Oligo Persistent: Oligoarticular JIA Persistent Variety, 5. Oligo extended: Oligoarticular JIA Extended Variety, 6. ERA: Enthesitis Related Arthritis.

Disease activities	Mean	SD	Range
Pain VAS (0 - 100 mm)	35.8	7.6	25 - 50
General VAS (0 - 100 mm)	25.2	6.7	20 - 40
Number of active joints	4.3	1.6	2 - 8
Number of limited joints	1.8	0.8	0 - 4

Table 2: *Distribution of patients by their disease activities at presentation (n = 52).*

Note: VAS: Visual Analog Scale.

6 months follow-up of the studied patient

Out of 52 who received treatment, only 12 patients arrived at our OPD for follow-up due to financial problem. Following are the data of their follow-up periods.

Disease	Number	Symptoms Free	Relapse
Poly RF - ve	7	7	0
Oligo persistent	3	2	1
Poly RF + ve	2	2	0

Although, treatment and follow-up were not part of this study, we provided NSAID (e.g. Ibuprofen, Naproxen) and oral prednisolone, sometimes intra-articular corticosteroid and intravenous pulsed methyl-prednisolone and methotrexate for the treatment of JIA. Follow-up was difficult because patients are not regular in the OPD due to financial constrain.

Discussion

The main purpose of our study was to describe the clinical features of children with JIA presented at our center as understanding the clinical characteristics of this disease and its subtypes in our country is essential to provide a better planning for medical care. In our study the mean ages at onset and presentation of the disease were 7.5 and 7.9 years respectively. In most of the studies [5,7-9] except the Turkey's study there is wide difference (a minimum of 3 years) between the age at onset and age at presentation, which might be due to delayed referral from the primary care physicians or financial constraint and/or lack of awareness on the part of the patients' guardians. But in our study, we find an average delay of only 4 months between the onset and first presentation at tertiary centres. It might be that there is no referral system in our health care setting and seeking specialized health care is at the discretion of the patients concerned. A male predominance was observed in our study, which is quite consistent with the findings of Indian and Turkey's studies [8,9], but sharply contrasts with those of Gutierrez-Suarez., *et al* [5]. A study conducted in South Africa by Weakley., *et al*. [17] however, showed no sex differential in the occurrence of JIA.

The present study shows that predominant subtype of JIA was RF negative polyarthritis (42.3%). Islam., *et al*. in a similar study in another tertiary hospital of Bangladesh also found polyarthritis JIA to be the highest. However, Shafiul and colleagues in a population-based study demonstrated oligoarthritis variety to be the highest (60%). The difference between the pattern of JIA between the previous population-based and the present hospital-based study might be that poly-arthritis cases being severe frequently seeks care from specialized hospital making its overrepresentation in hospital-based study. The occurrence of RF negative polyarthritis subtype in the present study is highest compared to all other relevant studies [5,7-9,17,18] conducted around the world. A limitation that must be mentioned in the present study is that the ILAR criteria require at least 2 positive RF assays to be done at least 3 months apart in the first 6 months of the disease in order to diagnose RF positive polyarthritis [1]. Due to resource constraints, obtaining of 2 RF titers is not standard practice in our setting, and one assay was considered sufficient to classify a patient with polyarthritis into positive or negative. This has been a difficulty with the ILAR classification, especially in poorly resourced settings. This difficulty is mentioned in the Indian study [8] and in a study of Nordic children [19]. Thus we believe that the RF negative polyarthritis patients may be overestimated in our polyarticular subtype as a child with one positive assay was classified as RF positive or negative polyarthritis without repeating the test for confirmation. The second most common arthritis in the present study was persistent oligoarthritis (25%). This is in line with the previously described data that showed that non-European populations have a decreased relative risk of suffering from oligoarthritis [20]. This may be a true reflection of a decreased prevalence or it may reflect that these patients have less obviously severe disease and would be underrepresented in a tertiary centre. The oligoarthritic children may do comparatively well in the community and may not feel to go to a higher centre for better treatment.

The prevalence of ERA was only 5.8%, which is much lower that found in Weakley's study (23%) and almost half than that reported in Turkish study (10.3%), but almost close to that reported in a recent UK longitudinal cohort study (7%) [6,9]. The Indian study however, so far reported the highest rate ERA (36.0%) [8]. The large PRINTO series states that there were too few a number of ERA patients, so they were excluded from further consideration [5]. The ERA has been described as being more prevalent in Asian populations including Indian populations [8,20]. It is less frequently observed in Black population. But it is more prevalent in the Western Cape region of South Africa, where a unique mix of population is found with a higher prevalence of colored people (44%) compared to Black people (34.9%) [21]. The colored population has mixed ancestry with some Asian and European heritage which may explain for the high levels of ERA. Very few patients were classified as systemic (7.7%) and extended oligoarthritis (7.7%). None had psoriatic or undifferentiated arthritis which may be a reflection of small sample size or hospital-based study.

Limitation of the Study

Our study is limited by sample size and hospital-based study. There are difficulties with the ILAR classification in our setting, specifically regarding the requirement of 2 rheumatoid factor tests to make the diagnosis confirm. Due to lack of presence in the follow-up schedule, the monitoring of the patient after receiving the treatment was difficult, therefore, the outcome of the disease was not properly quantified.

Conclusion

The present study revealed a different JIA profile compared to other international JIA studies. JIA was predominant in male children. RF negative polyarthritis is more common than has been described elsewhere. The second most common subtype was oligo-persistent. There is a lower rate of ERA.

Bibliography

1. Petty RE., *et al*. "International league of associations for rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton 2001". *The Journal of Rheumatology* 31 (2004): 390-392.

2. Hofer M and Southwood TR. "Classification of childhood arthritis". *Best Practice and Research: Clinical Rheumatology* 16 (2002): 379-389.

3. Martini A and Ruperto N. "Quality of life in juvenile idiopathic arthritis compared to healthy children". *Clinical and Experimental Rheumatology* 23.19 (2001): S1-S72.

4. Martini A and Ravelli A. "Juvenile idiopathic arthritis". *Lancet* 369 (2007): 767-776.

5. Gutierrez-Suarez R., *et al*. "Health-related quality of life of patients with juvenile idiopathic arthritis coming from 3 different geographic areas. The PRINTO multinational quality of life cohort study". *Rheumatology* 46.2 (2007): 314-320.

6. Hyrich K., *et al*. "Disease activity and disability in children with juvenile idiopathic arthritis one year following presentation to paediatric rheumatology. Results from childhood arthritis prospective study". *Rheumatology* 49 (2010): 116-122.

7. Amine B., *et al*. "Health related quality of life survey about children and adolescents with juvenile idiopathic arthritis". *Rheumatology International* 29 (2009): 275-279.

8. Kunjir V., *et al*. "Profile of indian patients with juvenile onset chronic inflammatory joint disease using the ILAR classification criteria for JIA: a community-based cohort study". India: s.n., vols. *The Journal of Rheumatology* 37.8 (2010): 1756-1762.

9. Yilmaz M., *et al*. "Juvenile idiopathic arthritis profile in Turkish children". *Pediatrics International* 50 (2008): 154-58.

10. Arendarczyk Z. "Rheumatoid arthritis in children up to the age of 15 in Poland (polish)". *Paediatric Policy* 52 (1977): 73-78.

11. Manners PJ and Diepeveen DA. "Prevalence of juvenile chronic arthritis in a population of 12-year-old children in urban Australia". *Pediatrics* 98 (1996): 84-90.

12. Sauremann RK., *et al*. "Epidemiology Juvenile Idiopathic Arthritis in a Multiethnic Cohort". *Arthritis and Rheumatism* 56.6 (2007): 1974-1984.

13. Kahn P. "Juvenile Idiopathic Arthritis: an update for the clinician". *Bulletin of the NYU Hospital for Joint Diseases* 70.3 (2012): 152-166.

14. Azam S., *et al*. "Prevalence and clinical pattern of juvenile idiopathic arthritis in a semi urban area of Bangladesh". *International Journal of Rheumatic Disease* 15 (2012): 116-120.

15. Islam MI., *et al*. "Pattern of Paediatric Rheumatic Diseases: An Experience in a Tertiary Care Hospital, Dhaka, Bangladesh". *Bangladesh Journal of Child Health* 37.2 (2013): 97-101.

16. Singh G., *et al*. "Measurement of health status in children with juvenile rheumatoid arthritis". *Arthritis and Rheumatology* 37 (1994): 1761-1769.

17. Weakley K., *et al*. "Juvenile idiopathic arthritis in two tertiary centers in the Western Cape, South Africa". *Pediatric Rheumatology* 10 (2012): 35.

18. Al-Hemairi MH., *et al*. "The Pattern of Juvenile Idiopathic Arthritis in a Single Tertiary Center in Saudi Arabia". *International Journal of Inflammation* 2015.2016 (2016): 8.

19. Berntson L., *et al*. "Construct Validity of ILAR and EULAR criteria in juvenile idiopathic arthritis: a population based incidence study from the Nordic countries". *The Journal of Rheumatology* 28 (2001): 2737-2743.

20. Sauremann RK., *et al*. "Epidemiology of juvenile idiopathic arthritis in a multiethnic cohort: Ethnicity as a risk factor". *Arthritis and Rheumatology* 56 (2007): 1974-1984.

21. Small K. "Demographic and socioeconomic trends for Cape town 1997-2006". City reports [Online] (2008).

Volume 8 Issue 12 December 2019