

A cross sectional study on juvenile idiopathic arthritis in paediatric population

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Abstract

Objective: To determine the frequency of common subtypes of juvenile idiopathic arthritis among paediatric population.

Methods: The cross-sectional study was conducted at the National Institute of Child Health, Karachi, from January to June, 2010, and comprised children below 16 years of age with arthritis in one or more joints for at least 6 weeks. Physical examination was done and detailed history was recorded. When >5 joints of the body were involved, it was considered polyarthritis. If <4 joints were involved, it was pauciarticular arthritis.

Results: Of the 95 cases in the study, 52(54.7%) were girls and 43(45.3%) were boys, with an overall mean age of 11±1.2 years. Polyarticular arthritis was found in 51(53.7%) subjects and pauciarticular arthritis in 44(46.3%). Out of 43 boys, 27(62.8%) had pauciarticular, while 35(67.3%) of the 52 girls had polyarticular arthritis.

Conclusion: Polyarticular arthritis was the most common sub-type. Proportion of polyarticular arthritis was high in female children, while pauciarticular arthritis was high in male children.

Keywords: JIA, Polyarticular arthritis, Pauciarticular arthritis. (JPMA 65: 370; 2015)

Introduction

Juvenile idiopathic arthritis (JIA) is a chronic idiopathic autoimmune disease with an annual incidence of 2 to 20 per 100,000 children and prevalence of JIA is estimated to be between 57 and 113 per 100,000 children.^{1,2} JIA is one of the most common chronic diseases of childhood, occurring as frequently as juvenile diabetes mellitus, four times more frequently than cystic fibrosis and sickle cell anaemia, and 10 times more frequently than acute lymphoblastic leukaemia, haemophilia, or muscular dystrophy.³ The three major subgroups distinguished during the first six months of arthritis are oligoarthritis (1 to 4 joints involved), polyarthritis (≥ 5 joints afflicted), and systemic onset JIA (with fever and papulomacular rash at the onset).⁴ The exact aetiology of JIA is unclear, but is considered to be the result of an immune dysregulation due to interactions between genetic and environment factors. No specific laboratory test can diagnose JIA. American College of Rheumatology (ACR) classification is the most widely accepted classification. It includes age less than 16 years, signs of arthritis in one or more joints, disease duration 6 weeks or longer, onset type defined in the first 6 months (i) polyarthritis: when 5 or more inflamed joints; (ii) oligoarthritis: when less than 5 joints and (iii) systemic onset disease: arthritis with characteristic fever and exclusion of other forms of juvenile arthritis.⁵

The burden and severity of arthritis and other rheumatic conditions continue to be a large and growing public health problem.⁶ In one of the studies conducted in Pakistan, polyarticular subtype was the commonest pattern seen in 51.6% cases while 44% of the patients had pauciarticular disease. Among these subtypes, pauciarticular is more prevalent in the West.⁷ Female gender and early age at onset are associated with poor functional outcome. Similarly, outcome also depends on JIA subtypes. Polyarticular JIA is associated with worse outcome in terms of functional impairment, delayed growth and disability.⁸

As local JIA data is scarce, it was a frequent presenting complaint in paediatric outpatient department (OPD). In this study, we identified the profile of JIA in terms of age at onset, gender and various subtypes of JIA in order to provide foundation for future studies.

Patients and Methods

The cross-sectional study was carried out at the National Institute of Child Health (NICH), Karachi, from January to June, 2010, and comprised children age <16 years with arthritis in one or more joints for at least 6 weeks. The sample size was calculated on the basis of $P = 44\%$,⁸ $d = 10\%$ and $1 - \alpha = 95\%$. Ethical approval was obtained from institutional committee and informed consent was taken from the parents concerned. Physical examination was done and detailed history was conducted. There were complaints of pain, inability to flex joints in the morning, warmth on inspection and swelling compared to other joints. When ≥ 5 joints of the body were involved, it was considered polyarthritis. If ≤ 4 joints were involved it was

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pauciarticular arthritis. Patients were classified into subtypes according to the ACR criteria. All this information was recorded and collected through a specially-designed proforma.

All analyses were conducted using SPSS 16. Descriptive analysis was done for demographics. Mean \pm standard deviation was computed for quantitative variables i.e.; age, age at onset, duration of disease. Frequency and percentage was computed for qualitative variables like gender and common JIA subtypes.

Results

Of the 95 children in the study, 52(54.7%) were girls and 43(45.3%) were boys, with the male:female ratio being 1:1.2. Overall mean age was 11 ± 1.2 years (range: 4-14

Table-1: Demographic characteristics.

| | Frequency | Percent |
|---------------------------------------|-----------|---------|
| Gender | | |
| Male | 43 | 45.3 |
| Female | 52 | 54.7 |
| Age | | |
| Mean \pm SD = 9.95 ± 2.4 years | | |
| < 5 | 3 | 3.1 |
| 5 - 9 | 26 | 27.4 |
| 10 - 14 | 66 | 69.5 |
| Age of Onset | | |
| Mean \pm SD = 7.6 ± 2.02 years | | |
| ≤ 5 | 12 | 12.6 |
| 6 - 10 | 81 | 85.3 |
| > 10 | 2 | 2.1 |
| Duration of Disease | | |
| Mean \pm SD = 2.31 ± 1.37 years | | |
| ≤ 1 | 29 | 30.5 |
| > 1 | 66 | 69.5 |

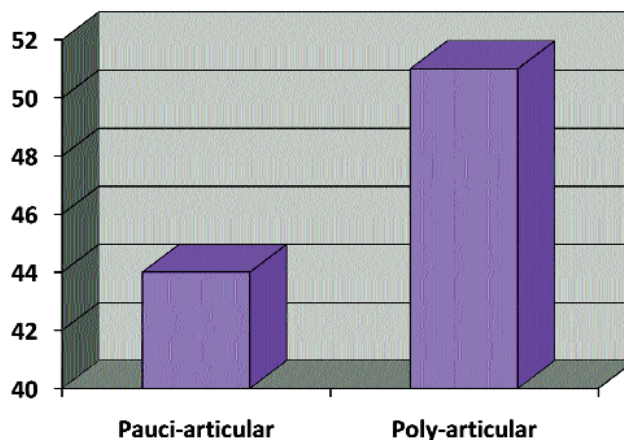
SD: Standard deviation.

Table-2: Common subtypes of juvenile idiopathic arthritis with respect to gender and duration of disease (n = 95).

| | Common sub types | | Total |
|------------------------------------|------------------|---------------|-------|
| | Pauciarticular | Polyarticular | |
| Gender | | | |
| Male | 27 (62.8%) | 16 (37.2%) | 43 |
| Female | 17 (32.7%) | 35 (67.3%) | 52 |
| Duration of disease (years) | | | |
| ≤ 1 | 16 (55.2%) | 13 (44.8%) | 29 |
| > 1 | 28 (42.4%) | 38 (57.6%) | 66 |

* Polyarticular juvenile idiopathic arthritis = Arthritis affecting > 5 joints in the first six month of disease.

* Pauciarticular juvenile idiopathic arthritis = Arthritis affecting < 4 joints in the first six months of disease.



* Polyarticular juvenile idiopathic arthritis = Arthritis affecting >5 joints in the first six month of disease.

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Figure: Common subtypes of juvenile idiopathic arthritis (n = 95).

years) (Table-1). The number of patients in the 10-14 years age group was 66(69.5%) followed by 26(27.4%) in the 5-9 bracket. Mean duration of disease was 2.31 ± 1.37 years. Duration of illness was more than 1 year in 66(69.5%) patients. Mean age at onset of disease was 7.6 ± 2.02 years.

Polyarticular arthritis was the most common subtype found in 51(53.7%) followed by pauciarticular arthritis in 44(46.3%) (Figure).

Out of 43 boys, 27(62.8%) had pauciarticular arthritis, while 35(67.3%) of the 52 girls had polyarticular arthritis. Proportion of pauciarticular arthritis was high in 16(55.2%) children who had arthritis ≤ 1 year, and proportion of polyarticular arthritis was high in 38(57.6%) children who had arthritis >1 years (Table-2).

Discussion

The term juvenile idiopathic arthritis is used for persistent joint inflammation or systemic illness with fever and rash of unknown cause lasting at least six weeks and starting in a child under 16 years of age. Several subgroups of JIA are well recognised, differing according to clinical manifestations, prognosis, specific autoimmune features and genetic determinants.

In Western societies, the annual incidence of JIA is 10-20/100000 in the paediatric population. Mono- and pauciarticular types of onset predominate, comprising about 50% to 75% patients. The proportion of polyarthritis is estimated to be 20% to 40%. Systemic onset JIA is less common; it is detected in 3% to 10%. The

variation in the proportions of the different subgroups depends in part on the study material: population- or clinic-based. Children with JIA are frequently positive for antinuclear antibodies (ANA).⁹

Patients with arthritis in five or more joints within the first 6 months of disease are diagnosed as having polyarticular JIA. This subtype includes children with rheumatoid factor (RF)-negative disease (20% to 30% of JIA patients) and RF-positive disease (5% to 10% of JIA patients).¹⁰ Both types affect girls more frequently than boys. RF-seronegative patients often develop polyarthritis in early childhood, which is in contrast with RF-seropositive patients who develop arthritis during late childhood and adolescence.

In our study polyarticular arthritis was the most common subtype found in 53.7% followed by pauciarticular arthritis in 46.3%.

In one study conducted in Pakistan, polyarticular subtype was the commonest pattern seen in 51.6% cases while 44% had pauciarticular disease. Among these subtypes, pauciarticular is more prevalent in the West.⁷ Female gender and early age at onset both are associated with poor functional outcome. Similarly, outcome also depends on JIA subtypes. Polyarticular JIA is associated with worse outcome in terms of functional impairment, delayed growth and disability.⁸

A study from Hong Kong reported that two most common subtypes were rheumatoid factor negative polyarticular (37%) and persistent oligoarticular JIA (24%). Most children presented at around 6 and 10 years of age.¹¹ One Indian study reported 28% oligoarthritis, 10% RF-positive polyarthritis and 20% RF-negative polyarthritis.¹²

A child who develops active arthritis of five or more joints after the first 6 months of disease is considered to have extended oligoarticular JIA. Up to 50% of oligoarticular patients may develop extended disease, and 30% will do so in the first 2 years after diagnosis. Risk factors for extended disease include ankle or wrist arthritis, hand disease, symmetric arthritis, arthritis of two to four joints, and an elevated erythrocyte sedimentation rate (ESR) and ANA titer.¹³ One study retrospectively evaluated JIA patients into adulthood with a median of 16.5 years of follow-up and found an overall remission rate of 12% in patients with extended oligoarticular JIA, compared with 75% in patients with persistent oligoarticular JIA.³

In this study, the proportion of polyarticular arthritis was high in girls than boys and pauciarticular was more in boys than girls, while males and females were earlier found to be equally affected by systemic JIA.¹⁴

In an Indian study there was a female preponderance in polyarthritis group, especially in RF-positive polyarthritis, where all five patients were girls. There was a male preponderance in the systemic and oligoarthritis groups.¹²

Proportion of pauciarticular arthritis was high in children who had arthritis ≤ 1 year, (52.2%) and proportion of Polyarticular arthritis was high in those who had arthritis >1 years (57.6%). Mean age of onset was 6.9 years in pauciarticular arthritis group and 7.1 years in polyarticular arthritis.

In an Indian study, 22% reported systemic-onset JIA, 28% pauciarticular and 30% polyarticular children.¹⁵

JIA is not a benign condition as some thought previously, but the attention it gets is disproportionately less poor. Prognostic indicators for patients with JIA are active systemic disease at 6 months, patients with polyarticular onset, RF positivity, persistent morning stiffness, tenosynovitis, subcutaneous nodules, or ANA. With better awareness of the significance of this condition and the continued efforts of prospective surveillance, it is hoped that more accurate data on prevalence and incidence could be obtained in the future. The new classification scheme for JIA may be a step forward in categorising patients with JIA, facilitating better communication in research, epidemiology and clinical care.

Conclusion

Timely diagnosis and early appropriate aggressive treatment of patients with poor prognostic features are indicated in order to improve quality of life and outcome. Large cohorts of patients from defined ethnic backgrounds are recommended.

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