Correspondence
Dr. Buddhi Prasad Paudyal,
Patan Hospital, Patan Academy
of Health Sciences, Lalitpur,
Nepal
Email:
buddhipaudyal@pahs.edu.np

Clinical profile of patients with juvenile idiopathic arthritis

Buddhi Prasad Paudyal,[1] Madhu Gyawalee[2]

[1]Associate Professor, Department of Medicine, [2]Lecturer, Department of Dermatology, Patan Hospital, Patan Academy of Health Sciences, Lalitpur, Nepal

ABSTRACT

Introductions: Juvenile idiopathic arthritis (JIA) is a heterogeneous group of disorders with different manifestations. There is lack of data locally. In this study, we describe the clinical profile and functional outcome of patients with JIA attending two rheumatology clinics in Lalitpur, Nepal.

Methods: This cross sectional study was conducted at Patan Hospital and Aarogya Health Home, Lalitpur, Nepal. The medical records of patients with juvenile arthritis during Jan 2013 to Dec 2015 were retrospectively reviewed for clinical profile and functional outcome of the disease.

Results: A total of 81 patients with arthritis, diagnosed before age 16 years, were seen during this period. Seventy-eight patients with JIA included 41 males and 37 females. Mean age of onset was 9.21 years. Polyarticular JIA was seen in 32 (42.2%), oligoarticular in 24 (30.7%), enthesitis-related in 13 (16.6%), and systemic-onset in 6 (7.7%) patients. Systemic complications were seen in 14 (17.9%) cases; five cases had uveitis; joint deformities were present in 28 (35.8%); sixty-seven (86%) children continued their education; functional limitation (Steinbrocker's functional class III and IV) was seen in 4 (5.1%). One child with oligoarticular JIA died.

Conclusions: The clinical profile of JIA in Nepalese patients is similar to studies from other parts of the world. During the short follow up, functional limitation was not a major problem.

Keywords: clinical profile, functional outcome, juvenile idiopathic arthritis

10

Journal of Patan Academy of Health Sciences. 2016 June;3(1):10-14.

INTRODUCTIONS

Juvenile idiopathic arthritis (JIA) is an umbrella-term describing a heterogeneous group of childhood conditions characterized by chronic arthritis beginning before the age of 16 years, persisting for at least 6 weeks, and having no other identifiable cause.[1] It is the most common rheumatic condition of childhood.[2] International League of Associations for Rheumatology (ILAR) has classified JIA into seven subtypes: systemic, oligoarticular, polyarticular rheumatoid factor (RF) positive and RF negative, enthesitis-related arthritis (ERA), psoriatic and "other" JIA.[1] It is an important cause of short- and long-term disability in children with decreased daily function and quality of life.[2]

Epidemiologic studies have noted wide differences in occurrence of JIA among different populations.[3] There is lack of published data on the epidemiologic pattern and clinical characteristics of JIA locally. This study provides information on the clinical profile of JIA from two rheumatology clinics from Lalitpur, Nepal.

METHODS

This was a cross sectional study conducted at Rheumatology Clinics of Patan Hospital and Aarogya Health Home, both located in Lalitpur district of Kathmnadu valley. Patan Hospital is a tertiary care teaching hospital of Patan Academy of Health Sciences (PAHS) whereas Aarogya Health Home is a private rheumatology clinic dedicated to the care of patients with rheumatic diseases. The medical records of children and youths diagnosed with JIA in the rheumatology services of these centres during January 2013 to December 2015 were retrospectively reviewed. Information about patient's demography and clinical details on subtype of JIA, age of onset, involved joints, systemic symptoms, systemic complications and presence of deformities were obtained from patient's clinical record sheet. Lab records included serological tests, RF, antinuclear antibody (ANA) and HLA B27. The hjILAR criteria requires at least two positive RF assays to be done at least three months apart in the first six months of the disease in order to diagnose RF positive polyarthritis,[1] due to cost factor only one positive or negative assay was considered sufficient to classify a patient with polyarthritis.

The impact of disease on patients' overall wellbeing was assessed by recording the Steinbrocker's[4] functional classes, continuation of education and mobility at last follow up. Steinbrocker's functional class is the physician's assessment of patients' capacity to perform activities of daily living, either at home or at work (Class I: normal functional status, Class II: adequate for normal activities, Class III: limited duties or self-care, Class IV: bed- or wheel-chair ridden, incapacitating condition). The death at home or other health facility was confirmed and recorded. Arthritis with a known cause (e.g., Enteropathic arthritis) or arthritis diagnosed past the age of 16 was excluded. Ethical approval was taken from Institutional Review Committee of PAHS. SPSS 16.0 was used for descriptive data analysis.

RESULTS

There were 81 JIA seen during the study period. Of these, three were excluded as diagnosis turned out to be different later (1 Enteropathic arthritis secondary to Crohn's disease, 1 Behcet's disease, and 1 Henoch-Schonlein purpura). The remaining 78 JIA were analysed, male 41 and female 37, male to female ratio 1.1:1. Polyarticular JIA were 33 (42.2%), 21 (26.9%) RF negative and 12 (15.3%) RF positive. Oligoarticular JIA were 24 (30.7%), ERA in 13 (16.6%) and systemic onset JIA in 6 (7.7%). Psoriatic and undifferentiated JIA were one (1.3%) each.

Complications
Systemic complications were seen in 14 (17.9%) patients and were more frequent in polyarticular JIA. Growth disturbances which included leg length discrepancy was seen in seven (9%) and retrognathism in one (1.3%) with longstanding RF positive polyarticular disease. Five (6.4%) had uveitis. One patient with RF positive polyarticular JIA had multiple complications: complete blindness due to bilateral uveitis and severe aortic regurgitation.

Joint deformities were present in 28 (35.8%) patients. Fixed flexion deformity, deformity of elbow and button hole deformity were found in 12 (15.4%), 5 (6.4%) and 1 (1.3%) respectively. Ten patients (12.8%) had multiple deformities in combination with fixed flexion deformities of elbows and knees.

Serological markers
Rheumatoid Factor was tested in all patients, ANA in 60 cases. The RF was positive in 12 (15.4%), all in polyarticular disease. Among the tested ones, ANA was positive in 3 (3.8%). The HLA B27 was done in 9 cases of ERA and 6 of these (66.7%) had this gene present.

Impact of disease on patient's wellbeing: functional outcome, education, mobility and death
Impact of disease on patients' overall wellbeing was assessed by Steinbrocker's functional class, continuation of education and mobility recorded on last follow up. There were 71 (90.9%) class I, 3 (3.8%) class II and 4 (5.1%) class III and nil in class IV. Seventy five (96%) were mobile and 3 (4%) crutch users had polyarticular disease. There were 11 (14%) given up study due to various reasons related to the disease and remaining 86% were continuing their education. One child (1.3%) with oligoarticular JIA died.

DISCUSSIONS

There was slight male dominance (53%) in our series similar to Indian[5] and Turkish[9] studies. This is in contrary to the studies from Western countries[7,8] with female predominance. The mean age in our series was 14.24 years, a little higher than other studies.[5-8] This is most likely due to the fact that we included all patients with JIA irrespective of current age who had developed the disease before the age of 16 years.

If RF status is not taken into consideration, polyarticular disease was the most commonly observed form of JIA (altogether 42.2% of which 26.9% were RF negative and 15.3% RF positive). However, oligoarticular JIA was the most frequent single entity observed. The distribution of all forms of JIA was more or less similar to the results from other studies.[5-8] One striking difference from neighbouring Indian study[5] was that RF negative polyarticular JIA and oligoarticular JIA were more common and ERA was less common in our series. Some of the children classified as RF negative polyarticular JIA may in reality be extended oligoarthritis as it was difficult to distinguish these two because of lack of routine follow ups and delay during the follow ups. Also, we did not subdivide oligoarthritis into persistent and extended forms as it was not possible to do so due to the retrospective nature of the study.

Due to limitation of resources, we obtained a single RF test to classify patients into RF positive or negative status, similar to other studies.[5,6] This might have caused over representation of RF positive polyarthritis patients in our series as a child with one positive assay was classified as RF positive polyarthritis without having a repeat test as suggested by ILAR. Only 3 (out of 60 tested), patients had positive ANA and all of these belonged to oligoarticular JIA. This low rate of ANA positivite as compared to Western studies could be due to the fact that most of the laboratories in our setting use ELISA as opposed to standard immunofluorescence method for this test.[10]

Systemic onset JIA was in similar frequency to studies from India and South Africa, though, much lesser than from Eastern Europe and Latin America.[5-8] The ERA, though less than that in India and South Africa, was a significant observation in our study. Other studies have also shown that ERA is more prevalent in Asian populations.[5, 11]

There was one case each of psoriatic and undifferentiated arthritis in our series. Psoriatic arthritis is not a common rheumatic disease in Nepalese population.[12] Our patient with psoriatic arthritis had family history of psoriasis in father. The small number of undifferentiated arthritis in our series could be due to small sample size.

Table 1. Comparison of JIA data from different countries/continents

Features	Present study	India [5]	South Africa [6]	Eastern Europe [7]	Latin America [8]
Number of Patients	78	235	78	668	397
Current age (years)	14.24	----	12.4	11.4	10.7
Age of onset (years)	9.21	12	7.3	6.8	6.6
Male (%)	52.6%	58%	50%	28%	32%
Polyarthritis	42.2%	29%	40.9%	33%	40.6%
RF positive polyarthritis	15.3%	12%	14%		
RF negative polyarthritis	26.9%	17%	26.9%		
Oligoarthritis	30.7%	21%	26.8%	43.6%	30.9%
Persistent oligoarthritis		17%	21.8%	31.6%	21.9%
Extended oligoarthritis		4%	5%	12%	9%
Systemic onset	7.7%	8%	7.69%	23%	28.5%
ERA	16.6%	36%	23%	---	---
Psoriatic arthritis	1.3%	1%	1.28%	---	---
Undifferentiated	1.3%	5%	0%	---	---

Note: JIA= Juvenile idiopathic arthritis , RF= rheumatoid factor, ERA=enthesitis-related arthritis

Systemic complications, particularly for polyarticular JIA was a major problem seen in our study with leg length shortening. This is due to the involvement of the knee joint and is reported commonly with longstanding JIA.[13] Uveitis was seen in 6.4%, less common than 14% reported from Germany.[13] Uveitis was most frequently observed in patients with ERA. One of our patients with RF positive polyarticular JIA had complete blindness due to bilateral uveitis, a dreadful complication of JIA. Joint deformities, mostly in the form of flexion deformities, were a common finding (35.8%) in our series, probably an effect of delayed initiation of treatment and lack of early physical therapy as children often develop flexion contractures if adequate physiotherapy is not instituted on time.

Although majority of patients with JIA have good physical outcome when they reach adulthood, many patients may experience functional limitation owing to joint deformities and destruction, growth abnormalities and retardation and osteoporosis.[14] Our patient follow up varied from few months to 2 years. During this follow up period, severe functional limitation (Steinbrocker's functional class III and IV) was seen in 5.1% of patients which is a very low figure probably due to short follow up. Western studies have shown that in longer follow up for 10-15 years, 30-56% patients have this degree of limitation.[14] Three patients with severe polyarticular disease in our series were crutch users. The high rate (86%) of continuation of education despite the debilitating nature of disease reflects that parents have given high priority for education of their children. The unexpected death of one child with well controlled oligoarticular JIA doesn't seem to be related to arthritis and may have some other cause.

Small sample size and clinic-based data in this study may have skewed the results in several ways. Patients attending our rheumatology clinics were the 'selected' ones and may have more severe disease than those in the community. Thus milder forms of JIA may not have been the part of this study as they have higher chances of going into remission and do not present to the specialist clinics. Owing to this, selection or referral bias, the information derived from this hospital based database like ours may exert some skewed impression on clinical characteristics (subtypes, complications and serological status) and functional outcome of the disease.

Despite these limitations, this study tries to fulfil the information gap on JIA from Nepal. More comprehensive research on larger sample size and longer follow up is needed.

CONCLUSIONS

Polyarticular disease was the most common form of JIA, followed by oligoarticular, enthesitis-related and systemic-onset JIAs. Systemic complications were frequent in polyarticular JIA with fixed flexion deformities of elbows and knees. Severe functional limitation was seen in few patients only.

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