

Study on clinico-serological profile of juvenile idiopathic arthritis in children

Das K¹, Swain A², Sahu S.K.³, Satpathy S.K.⁴

¹Dr. Kedarnath Das, Associate Professor, ²Dr. Arakhita Swain, Associate Professor, ³Dr. Sourava Kumar Sahu, Senior Resident, ⁴Dr. S.K. Satpathy, Professor, all authors are attached with Department of Pediatrics, SCB Medical College and S.V.P.P.G. Institute of Pediatrics, Cuttack, Odisha, India.

Address for correspondence: Dr. Kedarnath Das, Associate professor, Department of Paediatrics, SCB Medical College and S.V.P.P.G. Institute of Pediatrics, Cuttack-753002, Odisha. Phone- 9437385439, E-mail id dr.kedar2008@gmail.com

Abstract

Background: Chronic arthritis, the most common chronic rheumatic disease of childhood, is one of the more frequent chronic illnesses of children. Besides clinical examination, various hematological and immunological markers are being used to diagnose Juvenile Idiopathic Arthritis (JIA). The present study was designed to be conducted in our institution to find out the clinico-serological profile of JIA. **Materials and Methods:** All children under 14 years, attending the OPD and suspected to be suffering from Juvenile Idiopathic Arthritis, during September 2010 to September 2012 were included in the study. They were screened for the diagnosis of JIA. All the cases were subjected to hematological, immunological, biochemical and radiological investigation. Analysis was made on clinico-serological profile of the disease. **Results:** Majority out of 40 cases belonged to the age group 5-10 years and males out-numbered the females. Pallor was present in 100% of SOJIA and Polyarticular RF +ve JIA, 78.5% in Polyarticular RF -ve JIA, and 60% of Enthesitis related arthritis (ERA). In systemic onset JIA, soft tissue swelling was found in 66.67% cases. No cases of psoriatic arthritis or undifferentiated types were detected. Juxta articular osteoporosis was found in 66.67% cases. Methotrexate, Oral steroid and Hydroxychloroquine were the drugs used for most of the varieties of JIA. **Conclusion:** Even though, different varieties of JIA were detected, no cases of psoriatic arthritis or undifferentiated types were found. Chronic arthritis in childhood shows very diverse clinical pattern. JIA is like an iceberg, the diagnosis of which is usually missed due to lack of definite diagnostic parameters and less accessibility of the common man to the health facility.

Key words- Juvenile idiopathic arthritis (JIA), Rheumatoid arthritis, Soft tissue swelling, Narrowing of joint space, Joint erosion

Background

Chronic arthritis, the most common chronic rheumatic disease of childhood, is one of the more frequent chronic illnesses of children and an important cause of short and long term disability [1]. Its true incidence and prevalence are unknown [2]. The worldwide incidence of JIA ranges from 0.8 to 22.6/100,000 children per year, with prevalence ranges from 7 to 401/ 100, 000 [3]. The ILAR criteria have been applied for classification of JIA in clinical practice in various regions of the world: Eastern Meditterrean, India, Turkey, Italy and the UK to mention a few [4,5]. In India, Kunjir et al found enthesitis related arthritis

(ERA) to be the commonest subtype seen comprising 36% of JIA followed by Polyarthritis (29%), oligoarticular disease (21%), and systemic onset (8%) [6].

Besides clinical examination, various hematological and immunological markers are being used to diagnose JIA. A good number of cases are attending the OPD of our institution, S.C.B. Medical College & S.V.P .P.G.I.P, Cuttack. But there is paucity of studies and literature on JIA in Odisha in general and our institution in particular. Keeping all these factors in view the present study has been conducted at S.C.B. Medical College & S.V.P. P.G.I.P, Cuttack during a period extending from September, 2010 to September 2012.

Manuscript received: 26th April 2017
Reviewed: 04th May 2017
Author Corrected: 12th May 2017
Accepted for Publication: 19th May 2017

Aims and Objectives

1. To study the incidence of different subsets of JIA and its clinical profile in children.
2. To study the hematological change in different subset of JIA patients
3. To assess the immunological parameters like ANA, Rheumatoid arthritis

Factor, ASO titer, C-Reactive protein in different subset of JIA.

Materials & Methods

The hospital based, cross-sectional, prospective study was conducted in the department of Pediatrics of S.C.B. Medical College, and S.V.P.P.G.I.P, Cuttack for a period of 2 years extending from the month of September 2010 to September 2012. The study material comprised of all the children under the age of 14 years attending the OPD of SCB Medical College and SVPPGIP, Cuttack and suspected to be suffering from Juvenile Idiopathic Arthritis. After detailed history and physical examination, each joint was counted individually. Cervical spine, lumbar spine, thoracic spine, carpal joints of each hand, and tarsal joints of each foot were counted as 1 joint each. [6] Each

metacarpophalangeal, metatarsophalangeal, and proximal interphalangeal joint was counted as a separate joint. Enthesitis was defined by a history of moderately severe persistent pain and demonstration of tenderness at one or more bony insertion sites of ligaments/tendons/fascia, especially around the heel and knee region.

To exclude other known causes of arthritis and to evaluate the patient for diagnosis, complications and management the following investigations were done. All the patients were subjected to Hematological (Hb, DC, TLC, platelet count, ESR, CRP Reticulocyte count, BT, CT, peripheral smear), Immunological (ANA, RA factor, ASO, dsDNA), Biochemical (renal function test, liver function test) and Radiological (X ray of affected joint, chest and spine) investigations. Mantoux test was done to exclude TB arthritis.

For comparative study, data was analyzed for each of the JIA onset subtypes for age & sex distribution, duration of illness at the time of presentation, mode of onset of the disease, presence of prominent clinical features, frequency of involvement of individual joints of upper and lower extremities-unilaterally(asymmetrical) or bilaterally (symmetrical).

Results

This study of clinical & serological profile of Juvenile Idiopathic Arthritis was done in 40 pediatric patients diagnosed as per the International League of Association for Rheumatology (ILAR) criteria (1997). All the cases between the age groups of 0-14 years were included in the study. Out of 40 cases, 37 (92.5%) were from rural area and 03 (7.5%) were from urban area. Majority of cases belonged to the age group of 5-10 years followed by the age group between 10-14 years. The lowest and the highest age at which the disease manifested were 2 years and 14 years respectively. Different Clinical manifestations were joint pain and swelling(100%), morning stiffness (97.5%), limitation of joint movement (72.5%), tenderness of joints (75%), fever(47.5%), pallor (70%), loss of appetite (47.5%), lymphadenopathy (20%), hepatomegaly(25%), splenomegaly (10%) of the cases.

Uveitis was not noted in any case. Fever, rash, joint pain, pallor, hepatosplenomegaly, lymphadenopathy were the prominent sign and symptoms of systemic onset JIA. Pallor was present in 100% of systemic onset JIA, 36.36% in oligoarticular JIA, 78.5% in Polyarticular RF –ve JIA, 100% cases of Polyarticular RF +ve JIA and 60% of Enthesitis related arthritis(ERA). Loss of appetite was prominent in SOJIA, Polyarticular (both RF –ve & RF +ve) and was absent in oligoarticular & ERA variety (**Table-I**)

It was noted that Hb was <10 gm% in about 50% of cases of JIA and in 50% cases of systemic onset JIA, 64.29% cases of polyarticular RF-ve JIA, 100% cases of polyarticular RF +ve JIA. Hb was >10gm% in 81.82% cases of oligoarticular JIA and 60% cases of enthesitis related arthritis (ERA). (**Table-II**) ESR>20 mm in 1st hour in 100% cases of SOJIA, polyarticular RF +ve JIA and ERA, 81.82% cases of oligoarticular JIA, 85.71% of polyarticular RF-ve JIA. WBC count was >11,000 in all cases of SOJIA and polyarticular RF +ve JIA, 64.29% cases of polyarticular RF-ve JIA, 27.27% cases of oligoarticular JIA and 40% cases of ERA. **Platelet count** was above 2 lakh in 83.33% cases of SOJIA, 72.73% cases of oligoarticular JIA, 64.29% in polyarticular RF-ve JIA, 100% in polyarticular RF +ve JIA and 80% cases of ERA. **CRP** was positive in all cases of systemic onset JIA, polyarticular (RF +ve & RF –ve) and ERA.

Table- I: Clinical features in cases of JIA

Clinical Features	SOJIA	OJIA	RF-PJIA	RF +PJIA	ERA	PsA	UnA	TOTAL
Joint Pain	6	11	14	4	5	0	0	40 (100%)
Swelling	6	11	14	4	5	0	0	40 (100%)
Early Morning Stiffness	5	11	14	4	5	0	0	39 (97.5%)
Limitation Of Joint Movement	1	8	12	4	4	0	0	29
Tenderness Of Joint	4	5	13	3	5	0	0	30 (75%)
Loss Of Appetite	5	0	11	3	0	0	0	19
Pallor	6	4	11	4	3	0	0	28
Fever	6	4	3		2	0	0	19
Rash	2	0	0	0	0	0	0	2
Uveitis	0	0	0	0	0	0	0	0
Lymph Node Enlargement	6	0	1	0	1	0	0	8
Hepatomegaly	6	0	2	1	1	0	0	10
Splenomegaly	4	0	0	0	0	0	0	4
Sub Cutaneous Nodule	0	0	0	0	0	0	0	0
Erythema Nodosum	0	0	0	0	0	0	0	0
Pericardial Effusion	0	0	0	0	0	0	0	0

It was positive 72.73% cases of oligoarticular JIA. The positive ASO titre was noticed in 4 cases of polyarticular RF-ve JIA, 4 cases of polyarticular RF +ve JIA, 2 cases of oligoarticular JIA and in 1 case of systemic onset JIA.

This observation clearly shows that ESR is raised in all subsets of JIA and is maximum in cases of SOJIA. CRP is the only indicator which is very much raised in all subtypes of JIA. (Table- III).

Table-II: Hematological parameters in JIA cases

SUBTYPE	Hb (g%)		TLC (mm ³)		ESR(mm in 1 st hr)		TPC(mm ³)	
	<10	>10	<11000	>11000	<20	>20	<2lac	>2lac
SOJIA	3	3	0	6	0	6	1	5
OJIA	2	9	8	3	2	9	3	8
RF-PJIA	9	5	5	9	2	12	5	9
RF + PJIA	4	0	0	4	0	4	0	4
ERA	2	3	3	2	0	5	1	4
PsA	0	0	0	0	0	0	0	0
UnA	0	0	0	0	0	0	0	0
TOTAL	20	20	16	24	4	36	10	30

Table-III: ESR /CRP / ASO in cases of JIA

SUBTYPE	ESR(mm in 1 st hour)			CRP		ASO	
	Mean	SD	Range	Positive	Negative	Positive	Negative
SOJIA	93.67	34.77	55-135	6	0	1	5
OJIA	40.45	28.49	6-85	8	3	2	9
RF-PJIA	74.42	43.20	10-130	14	0	4	10
RF +PJIA	108.75	25.82	85-140	4	0	4	0
ERA	83	42.44	32-145	5	0	0	5
PsA	0	0	0	0	0	0	0
UnA	0	0	0	0	0	0	0

Rheumatoid factor was positive in 4 cases of polyarticular RF +ve JIA and was negative in all other cases. In the present study, ANA was positive in 2 cases of oligoarticular JIA and negative in all other cases of JIA. In these two ANA positive cases double stranded DNA test was done and it was found to be negative (**Table-IV**).

Soft tissue swelling was present in 38 cases, narrowing of joint space in 15 cases, erosion of joints in 4 cases and juxta articular osteoporosis in 8 cases. In SOJIA soft tissue swelling was found in 66.67% cases.

Juxta articular osteoporosis in 66.67% cases. Narrowing of joint space and joint erosion were not found.

In oligoarticular JIA, all cases had soft tissue swelling and narrowing of joint space was found in 54.55% of cases. Juxta articular osteoporosis was found in 81.82% cases. None had joint erosion.

Among polyarticular RF-ve JIA 14.29% cases have joint erosion and also juxta articular osteoporosis 35.71% cases have narrowing of joint space (**Table-V**).

Table-IV: Rheumatoid Factor and ANA status in cases of JIA

JIA Subtype	Rheumatoid Factor		ANA	
	Positive	Negative	Positive	Negative
SOJIA	0	6	0	6
OJIA	0	11	2	9
RF-PJIA	0	14	0	14
RF +PJIA	4	0	0	4
ERA	0	5	0	5
PsA	0	0	0	0
UnA	0	0	0	0

Table -V: Radiological changes in cases of JIA

JIA Subtype	STS		NJS		JE		JAO	
	+VE	-VE	+VE	-VE	+VE	-VE	+VE	-VE
SOJIA	4	2	0	6	0	6	4	2
OJIA	11	0	6	5	0	11	2	9
RF-PJIA	14	0	5	9	2	12	2	12
RF +PJIA	4	0	4	0	2	2	0	4
ERA	5	0	0	5	0	5	0	5
PsA	0	0	0	0	0	0	0	0
UnA	0	0	0	0	0	0	0	0

STS=Soft tissue swelling, NJS=Narrowing of joint space, JE=Joint erosion, JAO=Juxta articular osteoporosis

Discussion

This study of clinical & serological profile of Juvenile Idiopathic Arthritis comprised 40 pediatric patients diagnosed as per the International League of Association for Rheumatology (ILAR) criteria (1997) [4,5,6]. Here the cases between the age group 0-14 years were included because the children above 14 years are attending the Medicine OPD as per the rules of the hospital. Males out-numbered the females with a male to female ratio of 1: 0.54. Similar age and sex incidence has been reported by Seth V et al [7].

Joint pain and swelling (100%), morning stiffness (97.5%), limitation of joint movement (72.5%), tenderness of joints (75%), fever (47.5%), pallor (70%), loss of appetite (47.5%), lymphadenopathy (20%), hepatomegaly (25%), splenomegaly (10%) of the cases. Similar observations were found in other studies [8]. Fever, rash joint pain, pallor, hepatosplenomegaly, lymphadenopathy were the prominent sign and symptoms of systemic onset JIA. Pallor was prominent in all subtypes except in oligoarticular JIA. Joint pain, swelling and morning stiffness were the common manifestation of oligoarthritis. The common symptomatology of polyarthritis cases were pain and swelling of joints, morning stiffness, limitation of joint movement, tenderness of joints, loss of appetite and pallor. In ERA joint pain, swelling, morning stiffness, tenderness of joints were present in all cases.

Polyarticular (both RF +ve & RF-ve cases) suffered from different degree of malnutrition commonly. 2 cases of Polyarticular and 1 case of systemic onset variety had grade III malnutrition. Rheumatoid factor was positive in 4 cases of polyarticular RF +ve JIA and was negative in all other cases. In the present study, ANA was positive in 2 cases of oligoarticular JIA and negative in all other cases of JIA. In these two ANA positive cases double stranded DNA test was done and it was found to be negative. Soft tissue swelling was present in 38 cases, narrowing of joint space in 15 cases, erosion of joints in 4 cases and juxta articular osteoporosis in 8 cases.

In SOJIA, soft tissue swelling was found in 66.67% cases and Juxta articular osteoporosis in 66.67% cases. Narrowing of joint space and joint erosion were not found. Similar was the observation made by Cassidy JT et al [1]. In oligoarticular JIA, all cases had soft tissue swelling and narrowing of joint space was found in 54.55% of cases.

Juxta articular osteoporosis was found in 81.82% cases. None had joint erosion. Among polyarticular RF-ve JIA 14.29% cases have joint erosion and also juxta articular osteoporosis 35.71% cases have narrowing of joint space [1].

Methotrexate was given to all cases of polyarticular RF +ve JIA patients, 71.43% cases of polyarticular RF -ve JIA, 66.67% cases of systemic onset JIA, 60% cases of ERA patients [9,10,11,12]. Oral steroid was given to all (100%) polyarticular (both RF +ve & RF -ve), systemic onset JIA, ERA patients & 63.63% of oligoarticular patients [13,14]. Hydroxychloroquine was given to 33.33% of cases of SOJIA, 9.1% cases of oligoarticular variety, 28.57% cases of polyarticular RF -ve JIA & 50% cases of polyarticular RF +ve JIA.

Conclusion

JIA is not uncommon in children of this part of Odisha, majority belonging to the age group 5-10 years and males out-numbering the females. Even though, different varieties of JIA were detected, no cases of psoriatic arthritis or undifferentiated types were found. The disease is insidious in onset in 75% of cases and acute in rest 25% cases. Chronic arthritis in childhood shows very diverse clinical pattern. JIA is like an iceberg, the diagnosis of which is usually missed due to lack of definite diagnostic parameters and less accessibility of the common man to the health facility.

Funding: Nil, **Conflict of interest:** None initiated, **Perission from IRB:** Yes

References

1. Cassidy JT, Petty RE. Chronic arthritis in childhood. In: Cassidy JT, Petty RE, editors. Text Book of Pediatric Rheumatology. 6th ed. Philadelphia: Elsevier Saunders, 2011: 211–314.
2. David M. Siegel, MPH; Harry L. Gewanter, Juvenile idiopathic arthritis, American Academy of Pediatrics Textbook of Pediatric Care, 2009.
3. Eveline Y. Wu, Heather A. Van Mater, C. Eglia Rabinovich, Juvenile Idiopathic Arthritis, Nelson Textbook of Pediatrics, 19th ed, 2011: 329-339.
4. Demirkaya E, Ozen S, Bilginer Y, Ayaz NA, Makay BB, Unsal E, et al. The distribution of juvenile idiopathic arthritis in the eastern Mediterranean: results

from the registry of the Turkish Paediatric Rheumatology Association. *ClinExpRheumatol* 2011; 29: 111–6.

5. Stabile A, Avallone L, Compagnone A, Ansuini V, Bertoni B, Rigante D. Focus on juvenile idiopathic arthritis according to the 2001 Edmonton revised classification from the International League of Associations for Rheumatology: an Italian experience. *Eur Rev Med Pharmacol Sci*. 2006 Sep-Oct;10(5):229-34.

6. Kunjir V, Venugopalan A, Chopra A. Profile of Indian patients with juvenile onset chronic inflammatory joint disease using the ILAR classification criteria for JIA: a community-based cohort study. *J Rheumatol*. 2010 Aug 1;37(8):1756-62. doi: 10.3899/jrheum.090937. Epub 2010 Jun 1.

7. Seth V, Kabra SK, Semwal OP, Jain Y. Clinico-immunological profile in juvenile rheumatoid arthritis--an Indian experience. *Indian J Pediatr*. 1996 May-Jun;63(3):293-300.

8. Aggrawal A, Mishra R. Juvenile chronic arthritis in India: Is it different from that seen in Western countries? *Rheum Int* 1994; 14 ; 53-56.

9. Alsufyani K, Ortiz-Alvarez O, Cabral DA, et al. The role of subcutaneous administration of methotrexate in children with juvenile idiopathic arthritis who have

failed oral methotrexate. *J Rheumatol* 2004; 31(1): 179–82.

10. Hunt PG, Rose CD, McIlvain-Simpson G, Tejani S. The effects of daily intake of folic acid on the efficacy of methotrexate therapy in children with juvenile rheumatoid arthritis. A controlled study. *J Rheumatol*. 1997 Nov;24(11):2230-2.

11. Burgos-Vargas R, Vazquez-Mellado J, Pacheco-Tena C, et al. A 26 week randomised, double blind, placebo controlled exploratory study of sulfasalazine in juvenile onset spondyloarthropathies. *Ann Rheum Dis* 2002; 61: 941–2.

12. Ilowite NT. Current treatment of juvenile rheumatoid arthritis. *Pediatrics*. 2002 Jan;109(1):109-15.

13. Broström E, Hagelberg S, Haglund-Akerlind Y. Effect of joint injections in children with juvenile idiopathic arthritis: evaluation by 3D-gait analysis. *Acta Paediatr*. 2004 Jul;93(7):906-10.

14. Ruperto N, Murray KJ, Gerloni V, et al. Pediatric Rheumatology International Trials Organization. A randomized trial of parenteral methotrexate comparing an intermediate dose with a higher dose in children with juvenile idiopathic arthritis who failed to respond to standard doses of methotrexate. *Arthritis Rheum*. 2004 Jul;50(7):2191-201.

.....
How to cite this article?

Das K, Swain A, Sahu S.K, Satpathy S.K. Study on clinico-serological profile of juvenile idiopathic arthritis in children. *J PediatrRes*.2017;4(05):322-327.doi:10. 17511/ijpr.2017.i05.06
.....