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ORIGINAL ARTICLE

A STUDY OF PROFILE AND PATTERNS OF "JOINT INVOLVEMENT" IN UNDER 15 YEARS AGE GROUP CHILDREN WITH ACUTE RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE

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ABSTRACT: CONTEXT (BACKGROUND): Acute Rheumatic fever / Rheumatic Heart disease is the most common acquired childhood heart disease diagnosis made in India. Poly Arthritis is one of the common manifestations of the disease and making it one among many differential diagnoses for sub-acute arthritis. **AIMS & OBJECTIVES:** To study the profile and patterns of joint involvement in children with Acute Rheumatic fever / Rheumatic Heart disease. **MATERIALS & METHODS:** The Study was conducted in a tertiary care hospital for 2 years period on less than 15 years children diagnosed as Acute Rheumatic fever / Rheumatic Heart disease as per Jones criteria (Updated 1992). **RESULTS:** Joint involvement is one of the common manifestations of Acute Rheumatic fever / Rheumatic Heart disease. In our study Joint involvement is found in 50% cases (18 out of 36 cases) i.e., 87.5% (7 out of 8cases) of first attack cases and 39.28 %(11 out 28 cases) of reactivation cases. Further we found Polyarthritis is the predominant pattern of joint involvement. **CONCLUSION:** Basing on the results of our study we conclude that Joint involvement was observed in majority of cases, more so in first attack cases of Acute Rheumatic fever. Further Polyarthritis is the predominant pattern in our study. We found Knee Joint involvement as most common joint to be involved in children with Acute Rheumatic fever / Rheumatic Heart disease.

KEYWORDS: Acute Rheumatic fever, Rheumatic Heart disease, Polyarthritis, Arthralgia.

INTRODUCTION[1]: Rheumatic fever is a multi-systemic disease affecting primarily the heart, joints, brain, cutaneous and subcutaneous tissue secondary to an immune reaction to Group A Beta hemolytic streptococcal infection by rheumatogenic strains 1, 3, 5, 6, 18, and 24. Reactivation of Rheumatic fever of only by Group A streptococcal pharyngitis but not by any other infection or illness. Rheumatic fever is characterized by a number of clinical and laboratory abnormalities, the combination of which makes the diagnosis very probable.

The incidence of Rheumatic fever is very low in developed countries compared to developing countries. The group A Beta hemolytic streptococcus causes Rheumatic fever is a firmly established fact and though there is no experimental model to support this fact. Clinical, epidemiological immunological evidence is there to support this fact. Rheumatic heart disease constitutes from 16.5 to 50 percent of the cardiac patients in a hospital. The available data regarding prevalence rate for Rheumatic heart disease in India as follows: In the village population near Agra was 2/1000 and in the urban population of Chandigarh it was 2.07/1000 for women and 1.23/1000 for men. Similar figures were obtained in Delhi. A survey of 11 cities in India showed a prevalence rate of 0.55 to 0.67/1000. A survey conducted by the Indian council of Medical Research (ICMR) involving 133,000 children 6 to 16 years in age showed the incidence to be 5.3/1000.[2] this shows the magnitude of the problem in the country.

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Paucity of criteria to diagnose Acute Rheumatic fever led to dilemma which resulted in under diagnosis or over diagnosis and this prompted Ducket Jones from Boston to propose a set of criteria to diagnose Acute Rheumatic fever. He sub grouped these criteria into major and minor, neither of them having any prognostic connotation in the year 1944.[3] these original criteria did not differentiate between arthritis and arthralgia and erythema marginatum was classified as minor criteria. Subsequently, the original Jones criteria have been modified four times. The latest revision by American Heart association was made in 1992 and according to this, the guidelines.[4] are as follows:

1. **Major Manifestations:** Carditis, polyarthritis, chorea, erythema marginatum and subcutaneous nodules.
2. **Minor Manifestations:** These include clinical findings i.e. fever and arthralgia, laboratory findings i.e. elevated acute phase reactants namely erythrocyte sedimentation rate and C-reactive protein and prolonged PR interval.
3. Supportive evidence of antecedent group a streptococcal infection in the form of positive throat culture or rapid streptococcal antigen test or elevated / rising streptococcal antibody titer.

For making a diagnosis of Acute Rheumatic fever (Initial attack), two major or one major and two minor manifestations should be present along with supporting evidence of antecedent group A streptococcal infection.

In 1988 W.H.O has made certain recommendations wherein three special categories in which diagnosis of Rheumatic fever is accepted without 2 major or one major and 2 minor criteria. These are (1) chorea (2) insidious / late onset carditis with no other explanation (3) Rheumatic recurrence. For (1) and (2) evidence of Streptococcal infection is not required with 'chorea' being a diagnosis of exclusion. In patients with documented Rheumatic heart disease, the presence of one criterion as of fever, arthralgia or elevated acute phase reactants suggests presumptive diagnosis of recurrence with evidence of previous Streptococcal infection.

The division of criteria for diagnosis into major and minor is based on the specificity for Rheumatic fever rather than the frequency or severity of the manifestation.

The frequency of manifestation varies from study to study depending on patient selection whether initial attacks or recurrent attacks were included and on the changing pattern of Rheumatic fever. There has been a declining trend in developed countries which has not been observed in developing countries.

ARTHRITIS[5]: This is a very common manifestation with a prevalence of 70 – 75 percent of cases according to western literature. However, the figures from India indicate that arthritis is seen in 30 to 50 percent of patients. Its incidence increases with age. Onset is usually 2 weeks after onset of streptococcal pharyngitis and rarely 35 days after infection. To diagnose arthritis, the joint should be red, hot, swollen and tender. Characteristically it is a migratory Polyarthritis involving large joints of extremities but Mono articular Arthritis is not uncommon and 25% of patients described by Feinstein and Spagnuolo 39 had Mono articular Arthritis. Sites involved commonly are knee and ankle followed by shoulder, spine and small joints of the hand. Joint pain is exquisite with even the weight of a blanket being intolerable, but the severity of pain may vary. Joint effusions with polymorpho nuclear leucocytosis may occur. Swelling and edema of articular and peri articular structures with serous effusion into the joint space occurs but pannus formation and erosion does not occur. Cellular infiltration and engorged dilated veins are seen. At a later stage fibrinoid lesions with histiocytic

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granulomas may appear. Cartilage is not involved. The large joints are most often affected, but the inflammation is transitory, leaving no residua.

Arthritis is self-limited rarely lasting more than 2-3 weeks dramatically relieved within 24 hours of salicylate therapy with no joint deformity except for rare occurrence of Jaccoud's arthritis involving metacarpophalangeal joints in which subluxation of joints occur without erosion, narrowing or fusion of joint spaces leading to ulnar deviation of the hand. This is usually associated with aortic and mitral valve disease.

If the patient was given salicylates -containing analgesics, these signs of inflammation may be mild. The Arthritis responds dramatically to salicylate therapy if patients treated with salicylates (with documented therapeutic levels) do not improve in 48 hours, the diagnosis of Acute Rheumatic fever probably is incorrect.

ARTHRALGIA[5]: Refers to joint pain without objective changes of Arthritis. It must not be considered a minor manifestation when Arthritis is used as a major manifestation in making the diagnosis of Rheumatic fever

Differential Diagnoses for Joint manifestations of Rheumatic Fever:

1. Juvenile Idiopathic Arthritis (JIA) formerly known as Juvenile Rheumatoid Arthritis (JRA) is often misdiagnosed as Acute Rheumatic fever. The following findings suggest Juvenile Idiopathic arthritis rather than Acute Rheumatic fever. Involvement of peripheral small joints, symmetrical involvement of large joints without migratory arthritis pallor of the involved joints, a more indolent course, no evidence of preceding streptococcal infection, and the absence of prompt response to salicylate therapy within 24 to 48 hours.
2. Other collagen vascular diseases (SLE, mixed connective tissue disease)
3. Reactive Arthritis, including post streptococcal arthritis.
4. Serum sickness.
5. Infections arthritis (Such as gonococcal).
6. Virus –associated acute arthritis (Rubella, parvovirus, hepatitis B virus, herpes viruses enteroviruses), is much more common in adults.
7. Hematologic disorders, such as sickle cell anemia and leukemia, should be considered in the differential diagnosis.

MANAGEMENT[5]: (With particular attention given to Joint Manifestations of Acute Rheumatic fever/ Rheumatic Heart disease):

1. When Acute Rheumatic fever is suggested by history and physical examination, one should obtain the following laboratory studies: complete blood count, acute-phase reactants (ESR and CRP), throat culture, ASO titer (And a second antibody titer, particularly with chorea), chest radiographs, and ECG. Cardiology consultation is indicated to clarify whether there is cardiac involvement; two-dimensional Echocardiographic and Doppler studies are usually performed at that time.
2. When the diagnosis of acute rheumatic fever is confirmed, one must educate the patient and parents about the need to prevent subsequent streptococcal infection through continuous antibiotic prophylaxis.

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3. Benzathine penicillin G, 0.6 to 1.2 million units intramuscularly, is given to eradicate streptococci at 3 to 4 weeks intervals. In patients who are allergic to penicillin, erythromycin, 40 mg/kg per day in two to four doses for 10 days, may be substituted for penicillin.
4. Bed rest of varying duration is recommended. The duration depends on the type and severity of the manifestations and may range from 1 week (For isolated arthritis) to several weeks for severe carditis. The ESR is a helpful guide to the rheumatic activity and therefore to the duration of restriction of activities.
5. Anti-inflammatory or suppressive therapy with salicylates or steroids must not be started until a definite diagnosis is made. Early suppressive therapy may interfere with a definite diagnosis of Acute Rheumatic fever by suppressing full development of joint manifestations and suppressing acute-phase reactants.
6. Therapy with anti-inflammatory agents should be started as soon as Acute Rheumatic fever has been diagnosed:
    a) For mild to moderate Carditis, aspirin alone is recommended in a dose of 90 to 100 mg/kg per day in four to six divided doses. This dose is continued for 4 to 8 weeks, depending on the clinical response. After improvement, the therapy is withdrawn gradually over 4 to 6 weeks while monitoring acute-phase reactants.
    b) For Arthritis, aspirin therapy is continued for 2 weeks and gradually withdrawn over the following 2 to 3 weeks. Rapid resolution of joint symptoms with aspirin within 24 to 36 hours is supportive evidence of the Arthritis of Acute Rheumatic fever.
    c) Prednisone (2 mg/kg per day in four divided doses for 2 to 6 weeks) is indicated only in cases of severe Carditis.

CLINICAL COURSE OF JOINT MANIFESTATIONS / PROGNOSIS: Arthritis subsides within a few days to several weeks, even without treatment, and does not causes permanent damage.

AIMS & OBJECTIVES: To study the profile and patterns of "Joint involvement" in children under 15 years age group with Acute Rheumatic fever / Rheumatic heart disease presenting to King George Hospital.

MATERIALS & METHODS OF STUDY: 36 children of less than 15 years age group satisfying the Revised Jones criteria (1992) diagnosed as Acute Rheumatic fever /Rheumatic heart disease (Including 8 cases first attack and 28 Reactivation cases) from paediatric Medical wards (Both inpatients and outpatients) of King George Hospital, Visakhapatnam were included in the study with respect to the following:

1. Profile and patterns of Joint Involvement (i.e. Arthralgia, Arthritis, Polyarthritis, Mono arthritis and Joints like knees, Ankles involvement etc.) and
2. Comparison of profile of Joint Involvement with other studies.

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RESULTS:

Clinical Feature	1st attack(n=8)		Reactivation(n =28)	
	Number of cases	%	Number of cases	%
Joint involvement	7	87.5	11	39.28
Polyarthritis	5	62.5	7	25
Monoarthritis	-	-	-	-
Arthralgia	2	25	3	10.71

Table 1 : Profile of joint involvement in first attack and Reactivation cases of rheumatic fever

Joint involvement seen in87.5% of first attack cases and 39.28% of Reactivation cases. Among them 62.5% (5 cases) of first attack cases and 25% (7 cases) of Reactivation cases of Rheumatic fever had Polyarthritis. None of the cases had Mono arthritis. But Arthralgia was observed lesser frequency than Arthritis. 25% (2 cases) of first attack and 10.71% (3cases) of Reactivation cases had Arthralgia.

TOTAL NO. OF CASES: 36

Feature	NUMBER OF CASES	% CASES
Joint involvement	18	50
Polyarthritis	12	33.33
Mono arthritis	-	-
Arthralgia	5	13.89
Large joints	18	100
Knee	17	94.44
Ankle	15	83
Wrist	4	22.22
Elbow	1	5.6
Hip	-	-
Shoulder	-	-
Small joints	2	11.11
Metacarpophalangeal joint	2	11.11
Cervical	-	-
Metatarsal	-	-
Sternoclavicular	-	-

Table 2 : Joint Involvement (1st Attack and Reactivation Cases Included)

Joint involvement was noted in 50% (18 cases out of 36) cases, and this study shows large joints to be involved in all the 18 cases (100%). Commonest joint being knee (94.44%), followed by ankle (83%), wrist [22.22%] elbow (5.6%). Isolated small joint involvement was not seen. Small joints were involved in 11.11% of cases along with large joints.

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Joint involved	Nair et al study.[6] (1990) n=120	Present study n=36
	%	%
Knee	77.27	94.44
Ankle	14.77	83
Wrist	3.40	22.22
Elbow	2.27	5.6
Hip	2.27	-
Shoulder	-	-
Small joints	23	11.11

Table 3: Comparative Study of Joint Involvement

This table shows that large joints are more commonly involved than small joints, knee joints being most commonly involved in agreement with as in case of Nair et al study (1990)[6]. Small joints are also affected in Rheumatic fever both studies but for a lesser incidence (11.11%) in the present study.

SUMMARY:

- Joint involvement (87.5%) was noted to be more common in the first attack of Acute Rheumatic fever whereas joint involvement was observed in 39.28% of reactivation cases.
- Polyarthritis was noted in (62.5%) cases and poly arthralgia in (25%) of first attack cases. Reactivation of Rheumatic fever cases also showed joint involvement to be more common.
- Isolated small joint involvement was not seen in any of our cases.

CONCLUSION: Basing on the results of our study we conclude that Joint involvement was observed majority of cases, more so in first attack cases of Acute Rheumatic fever. Further Polyarthritis is the predominant pattern in our study. We found Knee Joint Involvement as most common joint to be involved in Acute Rheumatic fever / Rheumatic Heart disease.

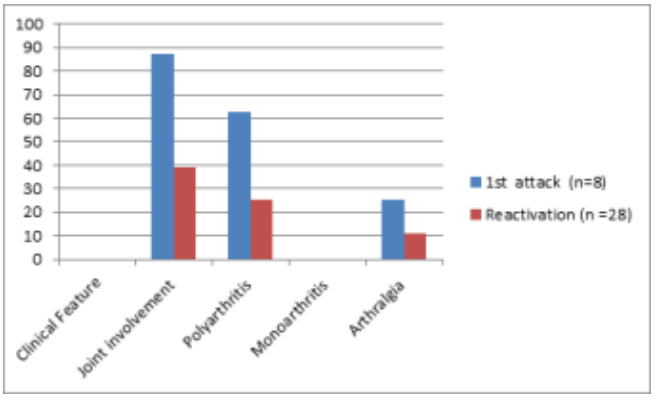

Fig. 1: Profile of joint involvement (%) in first attack and Reactivation cases of rheumatic fever

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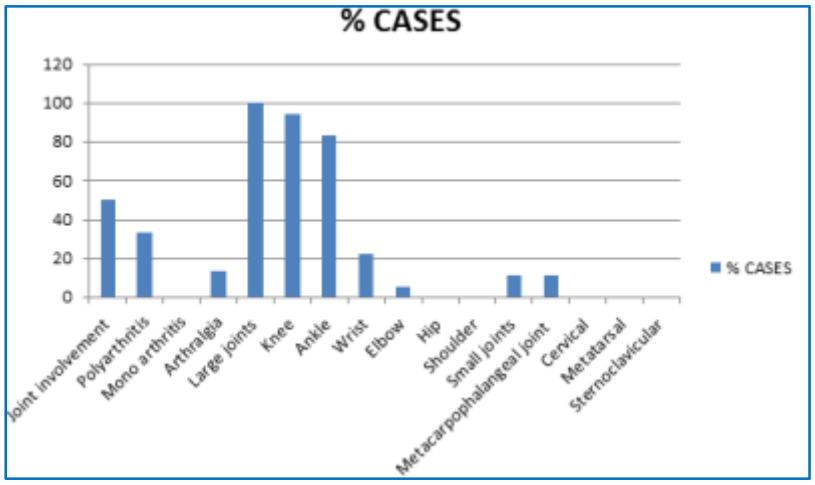

Fig. 2: Joint involvement (1st attack and reactivation cases included) (n=36)

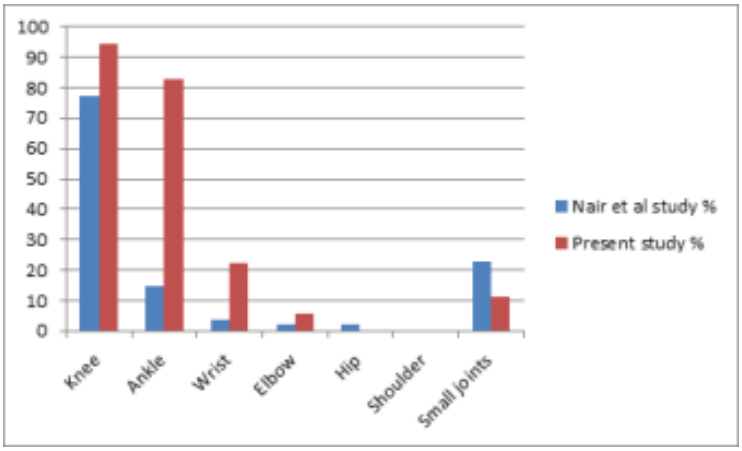

Fig. 3: Comparative Study of Joint Involvement

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