Indian Journal of Rheumatology 2012 May
Volume 7, Number 1 (Suppl); pp. 22–26

Review Article

Long-term outcome in juvenile idiopathic arthritis

Amita Aggarwal[1], Sajjan Shenoy[2], Ranjan Gupta[2]

ABSTRACT

Juvenile idiopathic arthritis is a chronic arthritis of childhood that continues in adulthood in about two-third children. In the past, it was associated with significant morbidity as reflected by physical disability, joint deformities, growth abnormalities, joint loss necessitating joint replacement at a young age along with decline in the psychosocial quality of life. In addition, most children took more time to complete education though they achieved their full potential in the end. In recent years, with better and early control of disease activity with the use of intra-articular steroids, methotrexate and biologics, the outcome has improved significantly with a hope that in future these children will have a near normal life.

Keywords: *Childhood arthritis, functional status, growth retardation, morbidity, uveitis*

Juvenile idiopathic arthritis (JIA) is the most common rheumatic disease of childhood that causes significant morbidity. Juvenile idiopathic arthritis is a heterogeneous group of disease with seven different categories. Thus, the long-term outcome is variable and depends on the category as well as on the pattern of joint involvement. Outcome in any chronic disease can be measured in multiple domains—the so-called 5Ds, disability, death, discomfort, dollar, and drug-related toxicity.[1] Disability in children is usually measured by childhood health assessment questionnaire (HAQ) and discomfort by using instruments that assess psychosocial, physical and mental impact of the disease such as the paediatric health rheumatology quality of life (PedHRQOL) scale. In children, another dimension is development and growth. Though, the outcome of JIA has improved significantly over the past two decades, it still causes significant disability. In this review, we will summarise the data on the long-term outcome in JIA in these various domains.

Disease activity persistence

In the past, there was the notion that JIA burns out and 75% children can expect to be free of inflammation in adulthood. This belief has now been challenged.[2–4] In fact, almost two-thirds of children continue to have active disease during adult hood with the proportion being higher in children with rheumatoid factor (RF) positive polyarticular category, a category where remission rates are lowest. In addition, a third of oligoarticular patients evolve into extended oligoarticular disease over time. Further, disease can relapse later in life even when children enter adulthood in remission. Almost 40% patients may have active disease after a follow-up period of 21 years.[5] The subsets with the worst prognosis (systemic onset JIA [SoJIA] and RF+ polyarthritis) form only 14% of JIA in paediatric age group but are 36.3% of the adult population with JIA.[5] Subclinical inflammation probably continues in even more patients as deformities continue to develop even in the absence of active synovitis. However, "burning out" may hold true for SoJIA as the level of clinical inflammation is lower in adults.[6]

Joint replacements

In the past, significant number of patients needed joint replacement, the common being the hip or the knee joint. Patients with SoJIA have the highest risk of joint failure

[1]Professor, [2]Senior Resident, Department of Clinical Immunology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India.
Correspondence: *Dr. Amita Aggarwal, email: amita@sgpgi.ac.in*
doi: 10.1016/S0973-3698(12)60025-9

whereas oligoarticular JIA and psoriatic arthritis have the least risk. Joint replacement especially in young adults with high demand of physical activity needs special considerations such as the choice of prostheses, cement/cementless procedure and need for redo surgery. Use of methotrexate has reduced the need for joint replacement[7] and this may even further reduce with the use of biologics.

Growth abnormalities

Mean heights of males and females with JIA remain shorter than the general population and standard deviations are increased indicating greater variation. Significant correlation has been observed between the length of time on steroids and the final height achieved. Though, the weights of these patients are similar to the general population, the standard deviations are increased.[8]

The growth abnormalities are predominantly seen in patients with prolonged inflammatory activity and use of steroids both of which occur often in children with SoJIA and polyarticular JIA.[8] In other categories, most children would have minimal growth abnormalities. In children with oligoarticular disease, involvement of unilateral joint of the lower limb can cause limb length discrepancy. In contrast to the leg length discrepancy in children, almost 82% patients with leg length discrepancy in adults have had hip prosthesis indicating that in adults with JIA, the main cause of leg length discrepancy is surgical.[6,7] Another localised growth defect is poor development of the mandible leading to micrognathia. This is due to the premature closure of the growing end of the temporomandibular joint arthritis. Almost 50% of children with micrognathia have SoJIA.[9,10]

Functional disability and quality of life

Early aggressive therapy has improved the outcome with the preservation of physical ability. Even though remission rates off medication are low in JIA, functional outcome is good with less than 10% children having severe disability.[11] In the past, 12% of patients had marked functional disability after 5 years of disease and nearly half after 16 years of disease.[12] Even in patients with functional independence, mild impairment in mobility, physical activity, household activity, depression, and health perception occurs.[13] Predictors of poor outcome include polyarticular disease, RF positivity, presence of radiological damage and involvement of the wrist and hip. Impact of the involvement of the elbows and shoulders on physical disability is higher than the involvement of hand joints.[14]

Education and unemployment

Though, patients with JIA attain similar levels of education, the time involved in the formal education is extended as a consequence of periods of ill health.[15] Good educational outcomes, irrespective of the functional disability, are likely to be due to the personality and coping strategies of patients and support from the family, teachers and schools.[16] However, in spite of good education, patients have significantly higher rates of unemployment. This has been attributed to poor self-esteem, poor perception of their health and low expectations by the patients and their family members. Vocational services and challenges may be delayed in these patients. There are problems in developing an "attitude and readiness for work" as well as issues of possible discrimination in the work place. Patients who were unemployed were more likely to be males, to have SoJIA or RF+ polyarticular JIA and to have greater disability.[5]

Psychosocial outcomes

Foster et al.[5] reported low mental summation and physical summation scores in all subtypes of JIA irrespective of their HAQ scores which suggests that the psychosocial impact of JIA is marked regardless of the degree of physical disability. Patients with oligoarticular and enthesitis-related arthritis (ERA) performed better than other categories. Using health-related quality of life instrument, Hovermanetal reported lower scores in all domains in patients with JIA.[17] In another study from Europe after nearly 18 years of disease, 22% had a general healthy quotient (GHQ)-30 score indicating psychiatric distress.[18] This had a significant correlation with pain and fatigue, indicating a relation to disease severity. However, patients with JIA are able to perform social activities similar to the controls.[15]

Fertility and obstetrical outcome

There appear to be no differences in marital status, live births and miscarriages. Child bearing is comparable but female patients are advised more frequently against pregnancies. This is unwarranted. Severity of the disease and the functional impairment are the limiting factors in decision-making for or against having children.[19]

Foetal and maternal outcomes of pregnancy are good. Mean birth weight was not significantly different from the general population. Seventy-four percent of the deliveries were normal vaginal deliveries. Of the 20 (26%) caesarean sections, JIA formed the basis for the operative delivery in