



Juvenile idiopathic arthritis

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Background

Chronic arthritis in childhood, called juvenile idiopathic arthritis (JIA), is a rare chronic condition of children and adolescents. Currently, there is no national surveillance of JIA in Canada. Although rarely fatal, the condition is long-term and associated with serious physical disability for many affected children. The disability associated with JIA has serious health impacts, such as pain, loss of independence, restrictions in daily activities and social participation as well as unemployment as young adults. An increased utilization of health care services has been demonstrated in addition to significant personal and societal costs.

Reliable and accurate data on the scope of chronic arthritis in children and adolescents in Canada is scarce. This information is crucial to determining the health services required by these individuals, and examining the gaps in health service provision. A limited number of epidemiologic studies have tried to measure the scope of JIA in Canada. Annual incidence rates have been reported between 5.3 and 10 per 100,000; and point prevalence estimates of 52 per 100,000 in Saskatchewan, and 32 per 100,000 in Manitoba have been calculated. The need for national data collection is illustrated by Malleson's study, which demonstrated wide confidence intervals for each province but narrower ones for the country. Furthermore, in two years only, 861 cases of juvenile arthritis of all types were reported; however, all these estimates were obtained from paediatric rheumatology specialty centres. The results obtained from primary care or community settings in other locales have always been much higher. Thus, although disease incidence is perceived as relatively low in Canada, there is the distinct possibility of significant underestimation of cases.



At present, there is no national data collection system that would enable surveillance of JIA in Canada. Previous attempts to collect data on children with JIA have been unsuccessful. The Canadian Paediatric Surveillance Program (CPSP) provides a unique opportunity to generate standardized surveillance data on JIA for the whole country, which could not be done otherwise. The Canadian Pediatric Rheumatology Research Group, a nationwide group of researchers dedicated to paediatric rheumatology research, is interested in collaborating with the CPSP to perform an effective national surveillance program for JIA. This project would interact with a concurrent new emerging team (NET) project, funded by the Canadian Institutes of Health Research (CIHR), in which 14 paediatric rheumatology centres in Canada are enrolling all newly diagnosed JIA patients into a long-term outcome cohort. A similar study is being carried out in the UK through the Welsh Paediatric Surveillance Unit (WPSU) and demonstrated that a surveillance program such as the CPSP will yield rigorous and accurate data on such rare conditions. A standardized approach to measure the scope and magnitude of JIA in Canada will facilitate development of appropriate interventions that ultimately can lead to improved quality of life for these children.

Methods

Through the established methodology of the CPSP, over 2,500 paediatricians and paediatric subspecialists will be actively surveyed for identified cases of JIA. The study will collect non-nominal, detailed case-specific data that will document the clinical features, epidemiological characteristics, familial autoimmune profile, and the current medical care practices provided to children with JIA. The 14 paediatric rheumatology centres (which encompasses nearly all paediatric rheumatologists in Canada) already involved in the research program ReACCh Out (Research on Arthritis in Canadian Children) will provide the data required for the CPSP study. The enrolment of newly diagnosed children with JIA seen in their clinics into a long-term outcome study will occur independent of the CPSP, and under no circumstances will pressure be applied to CPSP respondents to enlist patients for research.

Case definition

Report any child up to 16 years of age (up to 16th birthday) who presented for the first time with:

- Arthritis: persistent inflammation in one or more joints defined as
 - swelling or effusion, or
 - presence of two or more of the following signs:
 - limitation of range of motion
 - tenderness on motion
 - pain on motion
- Duration of disease: \geq six weeks

Exclusion criteria

All other relevant diseases (e.g., infection, malignancy, other systemic inflammatory diseases).

(The case definition is extracted from the definition of the International League of Associations for Rheumatology classification of Juvenile Idiopathic Arthritis, Petty et al, 2004 [see Appendix].)



Juvenile idiopathic arthritis (continued)

Objectives

- Ascertain the incidence of juvenile idiopathic arthritis in Canada.
- Determine feasibility and usefulness of an active surveillance system of juvenile idiopathic arthritis.
- Describe the demographics, including regional and ethnic variations of chronic childhood arthritis in Canada.
- Describe the clinical features of chronic childhood arthritis in Canada at presentation.
- Describe initial management strategies for chronic childhood arthritis in Canada, including treatment choices and referral strategies.
- Generate awareness of this rare disease among paediatric health care professionals.

Duration

October 2007 to September 2009

Expected number of cases

Based on the research team's previous experiences, it is expected that approximately 400 cases per year will be detected.

Ethical approval

Institutional Review Board, University of British Columbia
Research Review Board, BC Children's Hospital

Analysis and publication

The investigators will merge data and perform descriptive data analyses. Results will be disseminated by:

- Report provided to the Canadian Paediatric Rheumatology Research Group and the CPSP.
- Submission of an abstract to a rheumatology meeting and a paediatric meeting.
- Submission of a manuscript to a major rheumatology journal.
- Submission of a summary of the findings in a manuscript to a paediatric journal, to assist in increasing awareness amongst paediatricians of JIA.
- Communication of study results to families through paediatric rheumatology centres, and the website 'Just for Kids' sponsored by The Arthritis Society.
- Communication of results to the general public, government and research funding agencies through a surveillance report of JIA in Canada supported by the Public Health Agency of Canada (similar to the report of adult arthritis previously published [Health Canada, 2003]).

Bibliography

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Manners P, Bower C. Worldwide prevalence of juvenile arthritis – Why does it vary so much? *J Rheumatol* 2002;29:1520-30

Petty RE et al. International League of Associations for Rheumatology Classification for Juvenile Idiopathic Arthritis: Second Revision, Edmonton, 2001 *J Rheumatol* 2004;31:390-2

(Other references are available from the principal investigator or at the CPSP office.)



Appendix

Types of juvenile idiopathic arthritis

Systemic arthritis is defined as arthritis with a daily spiking fever for at least three days, or preceded by daily spiking fevers of at least two weeks’ duration, and the presence of at least one of (i) evanescent, non-fixed, erythematous rash; (ii) generalized lymphadenopathy; (iii) hepatomegaly or splenomegaly; or (iv) serositis. Other disorders with similar presentations should be excluded.

Oligoarthritis is defined as arthritis affecting one to four joints during the first six months of disease. If no additional joints become involved over time, this remains as persistent oligoarthritis; however, if five or more joints become affected after the first six months of disease, the term becomes extended oligoarthritis.

Polyarthritis affects five or more joints within the first six months of disease, and two forms are defined — rheumatoid factor (RF)-negative or RF-positive, in which case the RF must be positive on two occasions, at least three months apart, during the first six months of disease.

Psoriatic arthritis is defined as arthritis plus psoriasis, or arthritis and at least two of (i) dactylitis, (ii) family history of psoriasis in a first-degree relative, or (iii) nail pitting or onycholysis.

Enthesitis-related arthritis (ERA) is defined as (a) arthritis and enthesitis; or (b) arthritis or enthesitis with at least two of (i) sacroiliac joint tenderness and/or inflammatory spinal pain; (ii) presence of HLA B27; (iii) history in at least one first-degree relative of HLA B27-associated disease (ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease, Reiter’s syndrome or acute anterior uveitis); (iv) acute (symptomatic) anterior uveitis; or (v) onset of arthritis in a boy after age six.

Finally, a group defined as “*undifferentiated arthritis*” includes arthritis that either (i) does not fulfill criteria for any of the other categories, or (ii) fulfills criteria for more than one of the other categories.

In addition to these definitions, five separate exclusions have been listed as follows (see Table 1):

- a. Psoriasis or history in first-degree relative;
- b. Arthritis in a B27-positive male older than six years old;
- c. Ankylosing spondylitis, ERA, sacroiliitis with inflammatory bowel disease, Reiter’s syndrome or a history in a first-degree relative;
- d. RF positive on two occasions at least three months apart;
- e. Systemic arthritis.

Table 1: Exclusions for each category of juvenile rheumatoid arthritis

Arthritis type	Exclusions
Systemic	a, b, c, d
Oligoarthritis	a, b, c, d, e
Polyarthritis RF-negative	a, b, c, d, e
Polyarthritis RF-positive	a, b, c, e
Psoriatic arthritis	b, c, d, e
Enthesitis-related arthritis	a, d, e

Adapted from Petty RE et al (see *Bibliography*)