

¹ Centre Multisite Romand de Rhumatologie Pédiatrique, Départements universitaires de

Pédiatrie de Lausanne et Genève

² Kinderrheumatologie, Universitäts-Kinderspital, Zürich

³ Kinderrheumatologie, Universitäts-Kinderspital beider Basel, Basel

⁴ Poliklinik, Universitäts-Kinderklinik, Inselspital, Bern

Epidemiology of childhood arthritis in Switzerland

The authors are leading the pediatric rheumatology multidisciplinary clinics in the pediatric departments of the Swiss University Hospitals, Dr M. Hofer (Genève und Lausanne), Dr T. Saurenmann (Zürich), Dr D. Bolz (Basel) and Dr M.-J. Sauvain (Bern)

Chronic polyarthritis in children has been known since 1897, but has been given little attention until the last 20 years in paediatric rheumatology. There are now interest groups, specific scientific meetings. A European training programme in paediatric rheumatology is under preparation and quality standards have been defined for tertiary reference centres in paediatric rheumatology. Development of reference centres is dictated by the epidemiology of paediatric rheumatic diseases, the incidence of the disease, the proportion of severely affected children, and the long-term outcome. Such epidemiological data can be collected in registries of paediatric rheumatic diseases, as they already exist in the United Kingdom, France, and Germany.

In Switzerland the majority of the outpatient clinics for paediatric rheumatology are multidisciplinary clinics under the responsibility of both a paediatrician and a rheumatologist. Most of the physicians responsible for these clinics are members of the Swiss Working Group for Paediatric Rheumatology (SWGPR) which comprises paediatricians and rheumatologists with a special interest in paediatric rheumatology. Few years ago, the SWGPR has started to collect data on the patients seen by their members. But in the absence of technical and financial support, complete data collection was not possible. Paediatric rheumatologists in Switzerland may try to improve management of rheumatic diseases in childhood by conducting directly clinical studies, or by the participation to multicentric studies organised by a European organisation, PRINTO, the paediatric rheumatology international trials organisation.

In other specialties within paediatrics, epidemiologic studies have helped to understand the course of disease better and as consequence to increase the frequency of early and correct diagnosis and improve the therapeutic strategies.¹ In paediatric rheumatology, epidemiologic studies are difficult to conduct because of the multiple classifications used, the type of population studied, and the clinics where the patients are

examined (for example tertiary care centre or general hospital). Moreover, in children and adolescents with musculoskeletal pain, physicians are faced with a very large differential diagnosis, including infectious, inflammatory, hematological, mechanical, and orthopedical conditions. Beside more common diagnosis like infectious arthritis and Juvenile Idiopathic Arthritis (JIA), many rare diseases and syndromes may involve musculoskeletal tissues. In the presence of a non bacterial arthritis, parents are anxious to know its prognosis and evolution. Answers to these questions require epidemiological data. The current literature data is insufficient. In particular, it remains unclear what percentage of arthritis can be considered as post-infectious or will evolve as JIA with a more chronic course.

Disease classification

A major problem for the epidemiology of rheumatic diseases in children are the numerous and diverse classifications. In the past 20 years, the classifications and nomenclatures of the European League against Rheumatism (EULAR) and the American College of Rheumatology (ACR) have been primarily used; for example juvenile chronic arthritis (JCA) by EULAR and juvenile rheumatoid arthritis (JRA) by ACR. Both classifications are frequently interchanged although the diagnostic categories are not identical². Furthermore, criteria do not define homogeneous diagnostic categories. An international consensus on a classification for arthritis in childhood, and specifically of JIA, was proposed by the International League of Associations of Rheumatologists (ILAR) Task Force³. According to this classification, patients will be considered to have JIA if arthritis began before the 16th birthday, lasted for at least six weeks and if all other possible etiologies were excluded. After six months of disease based on precise inclusion and exclusion criteria, the patients will be

classified in one of the following categories: systemic arthritis, oligoarthritis (persistent or extended), rheumatoid factor negative polyarthritis, rheumatoid factor positive polyarthritis, enthesitis-related arthritis, psoriatic arthritis, and other arthritis (exact criteria reviewed in³). This last category re-groups all patients fitting in no other category and those fitting in more than one category. This new classification needs still to be validated with large groups of patients with JIA².

Disease prevalence and incidence

Epidemiologic studies have mainly come from Europe and North America, and have been scarce over the years. The published rates of incidence and prevalence showed wide ranges, depending on classifications used, inclusion criteria, length of the period studied and probably true population differences. In 1972, a first epidemiological study in France estimated the prevalence of chronic arthritis at eight per 100 000 children. Ten years later the estimated prevalence was 7.7 for Paris and 10 for Brittany per 100 000 children⁴. A recent Australian community-based study⁵, where 2241 school children were examined by a pediatric rheumatologist, showed a point prevalence of 400 per 100 000 children. The annual incidence in the French study⁴ was 1.3 for Paris and 1.9 per 100 000 children for Brittany. More recent studies in Sweden⁶ and in Finland⁷ reported higher incidences for JCA, 12 and 19.6 per 100 000/yr, respectively. An estimate of the incidence was also made with the Canadian registry and showed 4.1 per 100 000 children for juvenile arthritis, 0.3 for systemic lupus erythematosus and 0.15 for dermatomyositis⁸. Studies based on a registry probably underestimate the true annual incidence, because these rates do not include patients seen by physicians, who do not collaborate with the registry. For example, patients with dermatomyositis are also seen by paediatric neurologists and dermatologists. The highest rates were found in Scandinavia, where epidemiological studies are easier to perform because of the health care system.

Prognosis

Classically, it is stated "that up to 80% of children with chronic arthritis will be rid of inflammation when they reach adulthood". This statement has recently been questioned because

there is not enough supporting evidence. Like for other chronic diseases, the evaluation of the outcome of chronic arthritis in childhood is difficult because it depends on the classification used and the type of centre recruiting the patients. JIA may have a mild evolution without arthritis in adulthood and no remaining disability or have an aggressive course with multiple joint destruction and persisting disability in adulthood.

Functional outcome evaluated by Steinbrocker functional class⁹ has been assessed on patients with different length of follow-up. Severe functional limitations (Steinbrocker Functional Class III-IV) were found in 3% to 48%¹⁰ of the patients with juvenile arthritis with a follow-up of 3 to 37 years. Natural history of JIA was evaluated by Andersson Gäre et al. in a cohort study investigating the follow-up of 124 patients until the age of 16 years¹¹. At that age, 30% of the patients were in remission and 50% were still under medication. However, only 5% of the patients had severe functional limitations. Outcome is more favourable in population-based studies than in referral based settings, because of the different population selection. Thus, an epidemiological study based on a registry will give the necessary informations to provide health care and study on childhood population will determine the global outcome of paediatric rheumatic diseases.

Towards a Swiss registry

In Switzerland, until now there have been no community- or hospital-based epidemiological studies on paediatric rheumatic diseases. Thus, a national group of pediatricians has the project to study the incidence and prevalence, as well as the diagnostic distribution of arthralgia and arthritis on a large sample of the children population in Switzerland. They would like also to perform a pilot study for the establishment of a Swiss national registry for paediatric rheumatic diseases. This registry should evaluate the incidence, prevalence, diagnostic distribution, and long term outcome of childhood rheumatic diseases in Switzerland and help to recruit patients for clinical studies of diseases with few patients followed.

A better knowledge of the epidemiology of paediatric rheumatic diseases is essential to provide adequate care for these children and try to reduce poor outcome and long-term disability.

Michaël Hofer
Traudel Saurenmann
Dieter Bolz
Marie-Josèphe Sauvain

References

- 1 Kulig M, Bergmann R, Niggemann B, Burow G, Wahn U. Prediction of sensitization to inhalant allergens in childhood: evaluating family history, atopic dermatitis and sensitization to food allergens: the MAS Study Group Multicentre Allergy Study. *Clin Exp Allergy* 1998; 28: 1397–403.
- 2 Hofer M, Mouy R, Prieur AM. Juvenile idiopathic arthritides evaluated prospectively in a single center according to the Durban criteria. *J Rheumatol* 2001; 28: 1083–90.
- 3 Petty R, Southwood T, Baum J, Prieur AM. Revision of the proposed classification criteria for juvenile idiopathic arthritis: Durban 1997. *J Rheumatol* 1998; 25: 1991–4.
- 4 Le Gall E, Karman F, Blayo M, Prieur AM. Comparative epidemiologic study juvenile chronic arthritis in the western Paris area and Brittany. *Arch Pediatr* 1988; 35: 547–53.
- 5 Manners P, Diepeveen D. Prevalence of juvenile chronic arthritis in a population of 12-year-old children in urban Australia. *Pediatrics* 1996; 98: 84–90.
- 6 Andersson-Gäre B, Fasth A, Andersson J. Incidence and prevalence of juvenile chronic arthritis: a population survey. *Ann Rheum Dis* 1987; 46: 277–81.
- 7 Kunnamo I, Kallio P, Pelkonen P. Incidence of arthritis in urban Finnish children. *Arthritis Rheum* 1986; 29: 1232–8.
- 8 Malleson PN, Fung MY, Rosenberg AM. The incidence of paediatric rheumatic diseases: results from the canadian paediatric rheumatology association disease registry. *J Rheumatol* 1996; 23: 1981–7.
- 9 Steinbrocker O, Traeger CH, Batterman RC. Therapeutic criteria in rheumatoid arthritis. *JAMA* 1949; 140: 659–62.
- 10 Pedersen FK, Heilmann C, Friis J, Jørgensen B, Thomsen G. A follow-up investigation of 93 patients with juvenile chronic arthritis. *Ugeskr Laeger* 1987; 149: 2843–5.
- 11 Andersson-Gäre B, Fasth A. The natural history of juvenile chronic arthritis: a population based cohort study. Outcome. *J Rheumatol* 1995; 22: 308–19.