

Frequency of Uncommon Diseases: Is Juvenile Idiopathic Arthritis Underrecognized?



Accurate estimation of the frequency of rare or uncommon diseases is difficult. Some diseases, such as Kawasaki disease and Lyme disease, have only been relatively recently described, yet it is clear that examples of these disorders existed long before their definitive description evoked an epidemic of reports of similar cases. Reported frequencies of Kawasaki disease and Lyme disease vary enormously from place to place, reflecting at least in part differing genetic predispositions (in the case of Kawasaki disease) and causative agents (in the case of Lyme disease), but also differences in community awareness. Differences in the reported prevalence and incidence of idiopathic forms of childhood arthritis are also considerable, and more difficult to explain.

In this issue of *The Journal*, Manners and Bower address the problem of why reported prevalences and incidences of juvenile arthritis vary so much¹. This comprehensive survey of the published data indicates that the incidence of chronic arthritis of childhood ranges from 0.008 to 0.226 per 1000 children, and that the prevalence ranges from 0.07 to 4.01 per 1000 children. The authors point out several reasons for these differences: variation in diagnostic criteria, biases in ascertainment and differences in study design, low frequency of disease, and consequently, small numbers of patients in each study. The survey does not definitively answer the question it poses, but it provides a clear analysis of the existing data and emphasizes an important factor: the community based studies, although only 2 in number, yielded the highest disease prevalence. Does this mean that the children with chronic arthritis that are recognized in other reports are only those who have been able to make their way through the social and bureaucratic maze of medical care to be seen by a physician who recognizes the disease? In their community based study Manners and Diepeveen² reported a prevalence of chronic arthritis of 4.01 per 1000 children. To identify these patients the authors personally examined 2241 twelve-year-old school children in Western Australia. Significantly, most of the children with arthritis had not previously come to medical attention. It is unlikely that this is unique to Australia.

Innumerable children with chronic arthritis are diagnosed as having chronic sprain or growing pains, often reflecting that many physicians still fail to recognize that although uncommon, chronic idiopathic arthritis in children is not rare, whereas chronic sprain and growing pains causing joint swelling are!

Although differences in study design and available clinical expertise are almost certainly important contributors to the wide range in reported frequencies, we must not ignore the possibility that geography and ethnicity may contribute significantly to actual differences in the frequency of childhood arthritis. We know this to be the case for the HLA-B27 related arthritis. The data describing childhood arthritis from many parts of the world are so limited that firm conclusions about disease occurrence and the influence of genetics and geography cannot be made.

The interesting data from the Mayo Clinic^{3,4} suggest that in a single geographic area in the United States the incidence of chronic arthritis in children has decreased from 0.150 per 1000 between 1960 and 1969 to 0.141 between 1970 and 1979, and to 0.078 between 1980 and 1993. It is difficult to ascribe these changes to any known cause, but the data are unique, and similar studies from different areas of the world are needed to confirm or challenge this observation. If there is a world-wide trend to decreasing incidence of chronic childhood arthritis, this has implications not only for the interpretation of prevalence studies published as long ago as the 1950s, but also in planning and provision of health care and training of health professionals, and for determining the cause(s) of the disease(s).

This is the Bone and Joint Decade. It is an opportune time to grapple with this question: what is the global burden of chronic childhood arthritis? How can the lifelong impacts of a childhood disease be quantified? How can the effect of childhood arthritis on quality of life be meaningfully estimated in a world-wide population with differing expectations and socioeconomic circumstances? What is its influence on education, employability, fertility, lifespan? In their survey of existing data regarding prevalence and incidence of chronic arthritis in children, Manners and Bower

have provided a good starting point from which to approach these questions.

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