EDITORIALS

ON THE OCCURRENCE OF RHEUMATIC DISEASES IN THE UNITED KINGDOM

Knowledge of the occurrence of specific rheumatic diseases is of interest not only to epidemiologists but also to those planning clinical services, undertaking clinical research, and perhaps to the individual clinician in aiding diagnosis. Thus the recognition that young black females are at higher risk of SLE will increase the 'index of suspicion' when confronted with such a patient complaining of arthralgia. There are (contrary to the expectations of many inquirers to the ERU in Manchester) no routinely available data, in the United Kingdom or elsewhere, on the occurrence of the rheumatic disorders. Indeed, in the United Kingdom, morbidity data on incidence exist only for cancer (for example [1]) and for the notifiable infectious diseases (for example [2]).

There are data available from many disparate sources from which it is possible to build up a picture of the occurrence of the rheumatic diseases in the United Kingdom but it should be stressed there are many gaps. Mortality statistics are published annually by the Office of Populations Censuses and Surveys (for example [3]) which provide data by age, sex and area of residence. Mortality, however, is a poor proxy for incidence in rheumatology as very few of the conditions of interest have a sufficiently high or constant case-fatality rate. Data on hospital in-patients but not out-patients are available but, under the pre-Körner system of Hospital Activity Analysis (published annually as the Hospital In-Patient Enquiry [4]), the numerator is based on admission episodes rather than individuals; thus multiple admissions could not be identified separately. The United Kingdom is, however, well served by the Royal College of General Practitioners and its three National Morbidity Surveys. These studies, based on the continuous recording of every patient encounter during the study year by the (approximately) 50 participating general practices, have provided the only population-based record of prevalence and incidence of diseases [5–7]. There were no attempts at standardization of diagnoses and the reliability for the less common disorders might be low.

The other sources of data are published reports based on ad hoc population surveys and studies of hospital and general practice attenders with specific diagnoses. The population surveys of Leigh, Wensleydale and Watford [8] carried out in the 1950s and 1960s are unique in their scope and are still widely quoted. Changes in diagnostic classification and precision limit the contemporary applicability of these studies and, for the less common disorders, the samples studied were too small to provide reliable estimates. There have been numerous studies from other countries but their relevance to the United Kingdom is unclear. Particular mention should be made of the Rochester Epidemiology Project based at the Mayo Clinic. This latter institution, together with the Olmstead Medical Practice, with whom records are jointly accessible, provides the only source of medical care for the local community of Olmstead County in Minnesota. An enviable medical record system allows the retrospective recognition of all diagnosed cases with a disorder from this fixed population over a long time period. This data source has provided much of the known epidemiology of rheumatic disease for the USA (for example [9, 10]) and the denominator population, being of northern European origin, might have considerable similarity with the population in the United Kingdom.

It is relevant to emphasize the difference between prevalence (the rate of existing cases) and incidence (the rate of new cases arising in a specific time period, normally in one year). Both rates are normally expressed as a suitable multiple of 10 of the denominator, e.g. per 1000 or per 100,000 of the at-risk population, depending on the rarity of the disease. Generally incidence is a more useful measure. However, for diseases where dating the onset is practically impossible, for example osteoarthritis, prevalence is the more usual descriptor.

Studies of prevalence normally require a substantial population survey at a single point in time whereas incidence studies require the continuous monitoring of a population over a longer time period for the appearance of new cases. As the latter is frequently not feasible, surrogates such as the rate of new attenders to a medical care facility are frequently used.

In this issue of the Journal the first of a series of reviews is published covering the major rheumatic diseases. In these reviews the criteria in use have been emphasized as the speciality is dependent on arbitrary criteria for diagnosis, and changing fashions make interpretation of occurrence data difficult. The data sources mentioned above have been examined and the best estimates of occurrence in the United Kingdom for each of the conditions selected. Where they are available, in addition to the normal descriptors of age and sex, data are presented on geographic, racial and genetic differences in occurrence. The rate of hospital utilization and admission are also considered in order to provide the hospital clinician with some estimate of the proportion of the 'iceberg' seen in hospital. Finally, mortality and survival data are given where they contribute to estimating the community burden of the diseases. Colleagues in the United States recently published some prevalence estimates of the rheumatic diseases in that country [11] which were received with considerable interest based on the overwhelming number of reprint requests (R. Lawrence, personal communication). It is hoped that readers will find the
data presented in these reviews of interest and constructive suggestions regarding the content and presentation of the data will be welcomed.

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REFERENCES

THE PHENOMENON, SYNDROME AND DISEASE OF MAURICE RAYNAUD

Man is a tropical animal, less able to retain heat than to lose it. His neutral environmental temperature is 28°C and after a fall of only 8°C his metabolic rate must double or his body temperature will fall. Man’s response to cold, the putting on of clothes, permits him to live in temperate areas. Despite this, some people respond abnormally to cold. The best recognized of the cold related disorders is Raynaud’s phenomenon (RP).

It was in 1862 that Maurice Raynaud first defined his clinical syndrome as episodic digital ischaemia provoked by cold, cyanosis and emotion. It is classically manifest by pallor of the digits followed by cyanosis and rubor. The pallor reflects vasospasm, cyanosis the deoxygenation of static venous blood and the rubor reactive hyperaemia following the return of blood flow. RP affects 5–10% of the population and when severe can cause digital ulceration and gangrene [1]. Until recently, lack of knowledge in this area was reflected by the difficulties in the treatment and prediction of prognosis of RP. However, recent studies have investigated the extent of the vasospasm, its aetiology, progression and treatment and these ‘moving points’ in Raynaud’s research are outlined below.

One of the major problems is the question of terminology. In Europe, RP is the blanket term used to describe anyone suffering from cold-related digital vasospasm. RP is subdivided into secondary Raynaud’s syndrome (RS) where there is an associated disorder and primary Raynaud’s disease (RD) where there is not. This terminology has recently been ratified by the UK Raynaud’s Working Group. Unfortunately this European classification is not globally accepted and, additionally, other terms associated with the term ‘Raynaud’ are also used with different meanings by various authors resulting in a confusing overlap of synonyms. This confusion over nomenclature makes assessment of the literature difficult. Maricq [2] in her recent review concludes that the question of terminology will probably remain in a state of flux as long as the aetiology remains unknown and controversial. She stresses a most important point for all authors—all terminology attached to Raynaud’s name should be carefully defined in each scientific paper so that useful comparisons within the literature can be made.

Not only has the terminology used in RP been poorly defined but Raynaud’s original definition of RP is now known to require significant modification. For example, the full triphasic colour change is not now thought to be essential for diagnosis. Furthermore, in addition to the digits, the tips of the ear lobes, tongue and the nose can also be involved. A recent hypothesis suggesting more widespread vasospasm in this disorder has been postulated. Workers have detected decreased oesophageal [3] and myocardial perfusion [4] after cold challenge. Some workers have speculated that the kidney and lung lesions [5] seen in severe RS associated with systemic sclerosis (SS) may be accounted for in the same way. It is interesting to speculate that abnormalities of the vasculature may exist throughout the whole RP patient. These may contribute to the wide spectrum of symptomatologies seen in RP. A further modification of Raynaud’s definition is that other stimuli apart from cold and emotion can provoke an attack, for example, trauma, hormones [6] and chemicals such as...