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T. LEHNER and C. G. BARNES

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Foreword

Although the triple syndrome of aphthous ulceration, genital ulceration and iridocyclitis was clearly described by Hippocrates and recorded by several authors in the early part of this century, it is the Turkish dermatologist Hulüsi Behçet who has captured its eponymous title. Since his description in 1937, many other manifestations have been added to the clinical features of the syndrome, which now appear to overlap with disorders such as ulcerative colitis and Reiter's disease. It is generally agreed that the condition is sufficiently discrete to justify its specific identification as a disease rather than a syndrome.

This volume contains papers given at a recent international conference on Behçet's disease, arranged through the Royal Society of Medicine but held at the Royal College of Physicians of London. The prevalence of Behçet's disease is highly variable, ranging from less than 1 in 100,000 inhabitants of Yorkshire to 800 in 100,000 of the Japanese island of Hokkaido. An immunogenetic basis is suggested by distinct HLA associations. Discussions were dominated by the immunological abnormalities with which it is associated, leading inevitably to consideration of the possible role of viral infection as the triggering event in individuals thought to be immunogenetically susceptible.

This Conference, organized by Professor Thomas Lehner and Dr Colin Barnes, both of whom have made fundamental contributions to our knowledge of the disease, followed similar conferences held in Turkey in 1977 and Japan in 1981. The participation on this occasion of 200 representatives of 20 countries illustrates the growing interest in this unusual condition, with so many tantalising clues to its possible aetiology. That interest continues unabated is evidenced by the decision to hold a fourth meeting in the USA in 1989.

Professor Lehner and Dr Barnes are to be congratulated on having brought together so many expert clinicians and laboratory workers whose deliberations filled two fascinating days and whose manuscripts have now been brought together to provide the latest facts and concepts about this curious disorder.

Sir Raymond Hoffenberg KBE, MD, PhD
President, Royal College of Physicians of London
December 1985

Preface

A disease known to affect a variety of different tissues and organs attracts a corresponding multidisciplinary body of clinicians and researchers. This volume contains a fair review of the state of art presented and discussed at the 3rd International Conference on Behçet's Disease. The remarkable geographical distribution of this disease has been emphasized, by a global epidemiological assessment, with some clear conclusions. Although the disease is relatively common in the Middle East, the prevalence is highest in the Far East, but an increasing number of cases are reported from other countries.

The basic manifestations of oral and genital ulcers, with cutaneous and ocular disease, are associated with arthritic, neurological, vascular, intestinal and probably pulmonary and cartilagenous features. Communication between those involved in so many disciplines is often fraught with problems which this conference attempted to overcome.

The immunopathological basis for this intractable disease has been investigated in depth and has yielded valuable information. An immunogenetic basis has been widely accepted, with the association of an HLA class I gene product. The possibility of a viral aetiology has received some support, in that the presence of the herpes simplex virus genome has been confirmed in circulating mononuclear cells. Abnormal immune responses and manifestations, associated with the herpes simplex virus genome in specific immunoregulatory cells, might be responsible for some aspects of Behçet's disease. The enhanced thrombotic tendency has received a well deserved share of attention, with particular reference to platelet function and fibrinolytic activity.

Whilst there is no cure for this disease the benefits and limitations of drug treatment have been emphasized. This applies particularly to corticosteroids, azathioprine, chlorambucil and colchicine. The most important addition to the treatment of uveitis has been cyclosporin which is well documented by a double-masked clinical trial, its use in experimental uveitis and a thorough appreciation of its unwanted effects. Other methods of drug management are discussed but need scientific validation, as well as further ethical considerations.

The high degree of awareness of Behçet's disease outside the 'endemic' regions, aided by the ease of modern travel have been responsible for a remarkable worldwide interest in this disease. The international expertise embodied in this volume should serve as an ongoing reference until the next International Conference in 1989.

Colin G. Barnes *Thomas Lehner*

London, December 1985

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