HANDBOOK OF CLINICAL NEUROLOGY

VOLUME 35

Edited by

P.J. VINKEN and G.W. BRUYN

INFECTIONS OF THE NERVOUS SYSTEM

PART III

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in collaboration with

HAROLD L. KLAWANS

NORTH-HOLLAND PUBLISHING COMPANY

AMSTERDAM · NEW YORK · OXFORD

Library of Congress Catalog Card Number: 68-8297

ISBN for the complete set: 0 7204 7200 8
ISBN North-Holland for this volume: 0 7204 7235 0

195 illustrations, 54 tables

PUBLISHED BY:

ELSEVIER/NORTH-HOLLAND BIOMEDICAL PRESS
335 JAN VAN GALENSTRAAT, P. O. BOX 211
AMSTERDAM, THE NETHERLANDS

SOLE DISTRIBUTORS FOR THE U.S.A. AND CANADA:

ELSEVIER NORTH-HOLLAND, INC.
52 VANDERBILT AVENUE
NEW YORK, N. Y. 10017

PRINTED IN GERMANY

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Foreword to volumes 33, 34 and 35

The contents of these three volumes are fairly well defined by the title 'Infections of the Nervous System'. The contents of each volume are further defined by the decision to dedicate each volume to a separate class of etiologic agents. The further organization of each volume then posed its own particular questions.

The first volume was limited to infections caused by bacterial agents. The major issue to be decided was whether to organize this volume by etiologic agent or by type of infectious process. Other problems included which bacterial toxins to include and how much emphasis should be placed on the therapeutic aspects of each disease. After much discussion among the editors and many colleagues, especially Dr. Stuart Levin, it was decided to organize the volume primarily along etiological lines but to include separate chapters on specific types of infections which can be caused by numerous bacterial agents but in which the clinical characteristics of the syndrome are more dependent on the location of the infection than the causative agent. The chapters on brain abscesses and focal suppurative infections is an example of this approach. Bacterial meningitis is, of course, the most important class of such infections and is represented by both a general introductory chapter and a group of chapters on specific etiologic agents.

The decision to include a chapter on chronic arachnoiditis was not based on purely etiologic consideration but on the clinical consideration that this syndrome at times must be differentiated from bacterial infections of the linings of the brain and spinal cord.

Each year new antibiotics appear and the spectrum of bacterial responses to older antibiotics changes. Because of these two factors, up-to-date considerations on the pharmacologic therapy of bacterial infections can quickly become outdated and, it might be argued, should not be included in a Handbook of this nature. The fact remains, however, that we can medically treat many diseases but can actually cure only a few and of these most are infectious. To de-emphasize the most up-to-date pharmacologic approaches to these potentially curable but life threatening diseases could well be considered unjustified. We believe that despite the built in obsolescence, modern chemotherapy must be given its just due and major attempts have been made to ensure that the therapeutic aspects (both medical and pharmacologic, and, where applicable, surgical) are thoroughly and accurately presented.

Concerning bacterial toxins, those toxins which are the products of active infections within the body, e.g., diphtheria toxin and tetanus toxin, were included while bacterial toxins in which there is no actual infection, such as botulism, will be included in the volume on toxic agents.

The second volume is limited to viral and rickettsial diseases. It also includes a series of chapters on diseases of unknown etiology in which viruses or viral-like agents may play a role. Once again this volume includes both introductory general chapters as well as more specific chapters on particular etiologic agents. This field is one of great excitement including the work on slow virus infection which resulted in a Nobel Prize in 1976 for Carleton A. Gadjusek and perhaps even greater potential as the complex relationship between immunology and virology becomes better understood. Professor Richard Johnson, whose advice on the organization of this volume was most helpful, pointed out that it is often unclear where the field of virology ends and the field of immunology begins. Because of this a separate chapter on the immunologic aspects of viral infection has been included. This serves as an introduction to specific chapters on viral-induced syndromes with immunologic aspects and on the relationship of viruses to multiple sclerosis.

Many inflammatory and presumably infectious diseases of unknown etiology have been included in this volume. These include such topics as Behçet's disease, acute cerebellar ataxia, opsoclonus, Bannwarth's syndrome, acute hemorrhagic leukoencephalitis and chronic benign lymphocytic meningitis. The logic of including all of these chapters can, we are sure, be challenged since it is quite likely that neither viruses nor viral related immunologic processes will finally be implicated in all of these disorders. Since, in each case, one of these mechanisms remains a strong possibility, the discussion was made to include all of these disorders in this volume. Special recognition should be given to Dr. Robert M. McKendall for his help in organizing this volume.

The third and last volume includes diseases caused by all other classes of infectious organisms. In many ways this was the easiest of all the three volumes to organize since it is arranged entirely by etiologic agents. The complexity comes from the vast array of protozoa, helminths, and mycotic agents which are able to cause disease in man. Professor J. O. Trelles gave us valuable advice on the organization of this volume for which we remain indebted.

As always we are especially indebted to the editorial staff, in particular Brenda Vollers and Robert Stanley for their careful and thoughtful work which has helped to keep editorial delays and errors to an absolute minimum. We also recognize the contributions of Ms. Pat Gerdes and Ms. Genevieve Logan whose organization of the editorial work in Chicago was a major factor in the production of these volumes.

P. J. V.
G. W. B.
H. L. K.

Acknowledgement

Several illustrations and diagrams in this volume have been obtained from other publications. Some of the original figures have been slightly modified. In all cases reference is made to the original publications in the figure caption. The full sources can be found in the reference lists at the end of each chapter. The permission for the reproduction of this material is gratefully acknowledged.

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Contents

CHAPTER 1

Parasitic diseases and tropical neurology

J. O. TRELLES

Lima, Peru

From the time of Aristotle to the present, our knowledge of the living world has expanded to more than one million species of animals and 330,000 varieties of plants. Fortunately, few are able to become parasites and to cause disease in man, or to act as a vector of a parasite, but in view of this rich variety of life it is not surprising that such disease may take many different forms.

Parasitic disease of the nervous system is more common in the tropics and is sometimes regarded as synonymous with 'Tropical Neurology'. This is incorrect because many of the different types of these illnesses have a much wider distribution, and in the tropics other causes of ill-health are far more important, notably protein deficiency, in its commonest presentation as kwashiorkor.

The two aspects of general neurological disease and of parasitic infestation of the nervous system in the tropics overlap in many ways.

TROPICAL NEUROLOGY

The part of the earth bounded by the Tropics of Cancer and Capricorn, in which almost half the world's population lives, is characterized by a relatively static climate and stressful living conditions. Many infective agents flourish there because their vectors are prolific, and because factors that diminish natural resistance commonly affect the human inhabitants, for example severe climatic stress, nutritional deficiency and the many consequences of poverty and social deprivation.

However the majority of truly tropical diseases are dependent on the following ecological factors:

a) the presence of parasites; those that attack particularly the nervous system include trypanosomiasis, cysticercosis, neurobartonellosis, hydatid diseases, malaria and the mycoses; b) the presence of vectors that facilitate their transmission, although these are becoming less common because of the widespread use of insecticides, and c) the presence amongst the abundant jungle fauna of carriers of a number of virus diseases, some of which also attack the human nervous system.

Tropical conditions may also have a beneficial action in that the prevalence of certain grave neurological diseases is less than in temperate zones, for example the rarity of poliomyelitis epidemics and of multiple sclerosis.

The recent remarkable growth of communications and the development of tourism mean that visitors to the tropics from temperate zones may pick up a tropical infection of the nervous system, and so present a diagnostic problem on returning home. If only for this reason, although tropical neurology has recently become better known, it is important that it be reviewed in such a comprehensive textbook of neurology as the 'Handbook of Clinical Neurology'.

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Health in the tropics

Three factors must be kept in mind in any exact assessment of the effect of a tropical environment on the health of the population: 1) living conditions in the tropics, 2) working conditions in the tropics, and 3) the level of public health and governmental medical care.

The tropics and living conditions. Climate and environment have a considerable influence on the biology of individuals who live there, on their activity and life. This influence is much more marked on persons born in temperate zones who emigrate to the tropics, and was particularly obvious during the entire nineteenth century and the beginning of the twentieth when the tropics were colonized. Large numbers of immigrants, administrators and troops underwent a process of transplantation to the humid and rainy zones of Africa and Asia; most of these people came from Europe, with a prevalence of Anglo-Saxons, whereas the humid tropics of the Americas had been populated in previous centuries by European colonists mainly coming from Spain, Portugal and Italy. Some suffered intensely from the transfer to the tropics, where invigorating seasonal change is absent and the constant heat only varies in relative humidity, the latter increasing in the rainy season. The burning solar radiation can lead to madness and amok, so often described by writers; without adequate protection it can lead to heatstroke and sometimes to death, while it can injure the eyes and burn the poorly pigmented skin. The intense monotony is depressing and stressful and can lead to an attempt at flight, a search for an artificial paradise through hashish, opium, or absinthe, about which so many great writers, especially of the last century, have written masterpieces.

The same causes act on the native of the tropics but less intensely because centuries of living there have led to an adaptation process enabling man to survive and acclimatize himself to the most adverse conditions such as extreme heat with temperatures up to 50° C in some parts of the Sahara or on the banks of the Red Sea.

More important than the climate or the torrential rains is the diet, poor in protein and vitamins, that leads to conditions such as kwashiorkor or marasmus in children and malnutrition in adults. This undernutrition makes man easy prey to debilitating disease which renders him less productive.

In the tropics poverty, physical and biological destitution reign supreme. Only yesterday it was still common to see children with anaemic facies and emaciated bodies, their abdomen swollen with the enlarged liver and spleen of parasitic disease such as malaria, trypanosomiasis or kala azar. Hence the very high morbidity and mortality, especially in children.

Working conditions are unfavourable for both the immigrant and the native. Agricultural work is rendered tiring, exhausting and difficult by the tropical rains, the monsoons of Asia and the steamy heat of Amazonia, or by sheer heat in the absence of rain and by consequent dehydration.

Tropical stress factors have been studied and described by many authors; they serve to make comprehensible the adversity of working conditions for foreigner and native alike. It has been shown that working efficiency begins to fall off above 22° C and is severely impaired above 30° C. The heat profoundly affects physical activity and leads to a sensation of fatigue and apathy, diminishes sensory acuity, disturbs sleep and lowers appetite. All this is intensified by the humidity which leads to depression and boredom.

The prospect is made even more gloomy by the monotony of the climate, the lack of change, the swarms of insects and the constant fear of tropical diseases. Add to these the lack of culture, a propensity to alcoholism, and the ease with which drugs of addiction can be obtained and the picture is complete.

Public health and medical care facilities. In general, the state of public health and the means available to the various countries for health care are deficient.

Most of these countries are vast with a mainly rural population. They have an inadequate system of communications with few railway lines and few roads fit for motor transport; or there may be small communities on river banks linked only by water traffic. The communities are isolated, developing, far from a health care organization.

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Apart from a few modern cities, the population lives in deprived villages, places with inadequate environmental sanitation, in which infectious or parasitic diseases are readily conveyed by the water supply or irrigation system; others dwell in towns lacking an efficient water supply or drainage system and therefore favouring the spread of infection.

Spread of communicable disease is also favoured by social and cultural backwardness, illiteracy, and complete ignorance of the most elementary rules of hygiene and disease prevention.

The public health services and health care system are equally defective or inadequate, with scarcity of hospitals and health centres. The lack of medical and paramedical personnel, nurses, health inspectors, dentists, and so on, offers a clear explanation of the high morbidity rates still prevailing in some areas.

However, these adverse living and working conditions and this lack of welfare and security are slowly being improved, and in some areas they have been overcome thanks to modern technology with installation of air conditioning, more widespread use of refrigeration for food preservation, and health education. In addition, the authorities have in many countries made efforts with health campaigns in rural and urban areas to ensure satisfactory distribution of drinking water supplies and health and social services. More faculties of medicine have opened, special attention has been given in the curriculum to hygiene, preventive medicine and sanitation, and more doctors, nurses and sanitary inspectors have been trained. Medical care has been improved in urban areas and is organized in the countryside. Hence there is reason to hope that in time the situation will improve and the outlook cease to be so gloomy.

PARASITIC DISEASE OF THE NERVOUS SYSTEM

Parasites are any living organisms, plant or animal, which during a part or the whole of their existence live at the expense of other living organisms. Because every disease is the result of conflict between the pathogenic agent and the defense reactions of the host, better understanding of illness requires study of both these aspects.

Biological studies of parasites have revealed much about habitat, behaviour, nutrition, their reproductive mechanisms and mode of development, and the effect on them of the environment. As a result, the reasons for their present geographical occurrence have become understood and their possible spread can be predicted, thus permitting defensive and preventive measures aimed at their eradication.

The biological properties of parasites may also aid understanding of the mechanisms underlying the diseases that they cause, which arise by a variety of actions – mechanical, irritative, traumatic and destructive, invasive, toxic and infective. The host may have a varied effect on the growth and size of the parasites, and on their development; in some cases, it may neutralize or destroy them by immunological and other reactions.

The pathogenic action of a parasite varies with its genus. Protozoa, metazoa and fungi are the commonest types to affect the nervous system; arthropods act indirectly as their vectors. The pathogenic effect of a parasite depends on its species and on factors that control the degree of virulence within each species, as well as on the responses and reactions induced in the affected host. Clinical disease ensues only when these conditions favour development and growth of the parasite. Certain climatic or geographical conditions, such as a particular temperature and humidity will favour establishment of the disease and its vectors and its persistence in endemic or epidemic form.

Certain individual factors appear to promote or discourage parasitic infestation. Thus, certain races show high resistance to a number of parasitic diseases, a fact probably explicable by natural selection; children having less resistance are more prone to infection. The customs of certain religions help to combat disease by dietary prohibitions, for example the Moslem and Jewish faiths through their prohibition on eating pork have made cerebral cysticercosis a rarity in their communities. Other customs, however, may favour infestation; e.g. Proctor (1963) has suggested that the witch doctor cult in Rhodesia is one pathway for cysticercal infestation.

Although much has been discovered about particular diseases and their pathological ecology,

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scientific interest in tropical neurology, and study of the relevant parasitic diseases, is barely more than a century old (Spillane 1973) and much still remains to be discovered. It is important, however, not to repair a new syndrome or disorder until the classical literature has been thoroughly studied, and the possibility of a novel manifestation of a well-known condition excluded.

HISTORY

Knowledge has been acquired in a series of stages. The most remote was recognition of a few diseases caused by the larger parasites. Then much later identification of numerous other parasites began, first those visible to the naked eye, concomitantly with recognition of the diseases that they caused. After Pasteur began the characterisation of microscopic parasites and study of their diseases and finally, with discovery of their role in the transmission of infections, has come detailed examination of parasitic vectors, and use of our new understanding to combat and control these illnesses.

The most important landmarks in the history of parasitology extend throughout the written history of mankind. In the Ebers papyrus of 1600 B.C., the larger worms were described as possible human pathogens and in the following centuries the ancient Israelites and the Greeks and Romans were all familiar with various manifestations of cysticercosis and hydatid disease. The discovery of Fasciola hepatica in sheep by Johan de Brie in 1370 probably qualifies him as the first parasitologist. In 1550, Paranoli and, in 1558, Rumler mentioned human cysticercosis, and its parasitic nature was established by Malpighi in 1686. Further accounts of parasitic worms were published in the eighteenth century, but significant advances were delayed until the advent of the experimental method.

After Payet had identified Trichinella spiralis in 1838, increased attention was paid to diseases due to helminths. New species were described, such as Ankylostoma duodenalis in 1838 by Dubini, and Schistosoma haematobium in 1851 by Bilharz. The life cycles of Taenia solum, Echinococcus, Fasciola hepatica and Ankylostoma were established in the second half of the nineteenth century, and this led to the study of vectors of infective processes.

In 1687 Binomo and Cestoni discovered that a skin disease, 'the itch', was produced by a mite, Sarcoptes scabiei, and in 1834 Bassi showed that muscardine, a disease of silkworms, was due to a fungus. Between 1841 and 1844, Gauby found that human ringworm and thrush were also fungal diseases, and in this way, the foundation of mycology aided recognition of parasitology and parasitic diseases.

As regards protozoa, in 1859 Lambi discovered Giardia intestinalis, a flagellate protozoon, since called Lamblia in his honour. Lösch of St. Petersburg described Amoeba histolytica in 1875, and showed experimentally its pathogenic role in dysentery. Not until 1912 did Von Prowazek discover Acanthamoeba hartmanni, the cause of a primary meningo-encephalitis, the severity of which was not recognized until the studies of Culbertson et al. in 1959, and Butt in 1966; similar variety, Naegleria fowleri identified by Carter in 1966.

As regards blood parasites, the agents of malaria were identified by Laveran in 1880 (Plasmodium malariae), Grassi in 1890 (Plasmodium vivax) and by Welch in 1897 (Plasmodium falciparum). The sexual cycle, sporogony, was demonstrated by Ross in 1898. In 1899, Bruce discovered a trypanosome in cattle, and its role as the agent of sleeping sickness was recognized in 1901 by Dutton in Gambia. The Rhodesian trypanosome was discovered in 1910 by Stephen and Fantham, and the agent of American trypanosomiasis was recognized by Chagas in 1909. The agent of Carrion's disease, with its stages of Oroya fever and verruca peruviana, was identified by Barton in 1905. Other important flagellate protozoa that cause major diseases include the agent of oriental sore or cutaneous leishmaniasis, which was discovered by Borowsky in 1889, and was properly described as Leishmania tropica by Wright in 1903; the agent of visceral leishmaniasis or kala azar, described almost simultaneously by Leishman, Donovan, Laveran and Mesnil in 1903, for which reason it is known as Leishmania donovani; and the agent of mucocutaneous leishmaniasis (uhta), a South American variety, identified in 1908 by Lindenberg in Bahia. Lastly, another very important protozoon, the agent of

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