Narcolepsy and Hypersomnia

Narcolepsy and
Hypersomnia

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40 figures and 50 tables, 1980

S. Karger. Basel· München· Paris· London· New York· Sydney

This book is published with the arrangement of
AVICENUM, Czechoslovak Medical Press, Prague

National Library of Medicine, Cataloging in Publication
Roth, Bedri'ich
Narcolepsy and hypersomnia / Eedi'ich Roth; rev. and edited by Roger
WM 188 R845na
ISBN 3-8055-0490-X

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ISBN 3-8055-0490-X.

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Chapter 23. The "insufficiency of wakefulness" syndrome [subwakefulness

It is a very great pleasure to introduce Dr. Bedfich Roth's second volume on narcolepsy and the hypersomnias. Working as he does, quietly and carefully, at the Neurological Clinic of the renowned Charles University in Prague, Czechoslovakia, Dr. Roth has built up since 1949 a unique and very large series of patients with neurological sleep disorders. The frequent follow-up duration of 25 years or more in these patients, along with his talents as a clinical neurologist, give Dr. Roth an experience with such issues as prognosis, side-effects of chronic medication and other aspects of these conditions which to my knowledge is unique. Anyone who has had the good fortune to visit Dr. Roth in his clinic can attest to the completeness of his files on his patients.

The first volume of Dr. Roth was based upon a series of 251 patients (155 with various forms of narcolepsy, 96 with hypersomnia). It was published in 1957 in Czech, translated in 1962 into German and, unfortunately, never was made available in English. To give the reader a historical perspective, the only workers in the 1950s with a perhaps comparable series of well-studied patients were Daly and Yoss at the Mayo Clinic; the first publication of altered sleep/waking patterns in narcoleptics using modern polygraphic techniques was that of Rechtschaffen and co-Haborators in 1963; and most individuals other than Dr. Roth, who are presently active in the investigation of these interesting conditions, date the start of their activities to the early or mid 1960s or later. Indeed, so far as I know, the next book in the literature treating these diseases exclusively was not published until 1976.

The present volume brings Dr. Roth's experience up to date, now with a remarkable total of 620 patients - 360 with narcolepsy, 260 with hypersomnia. As well as providing a careful analysis of the symptomatology, etiology, laboratory findings (including sleep polygraphy of all patients examined since 1965) and other aspects of his patient series, the volume emphasizes areas in which Dr. Roth and his colleagues have been particularly
active. These include the classification of the numerous forms of narcolepsy and the hypersomnias, their genetic bases, the various patterns of abnormal drowsiness seen in daytime diagnostic EEGs, the altered sleep/waking patterns recorded by polygraphic techniques, the phenomenon of sleep drunkenness, and others.

It has been an honour to help Bedfieh Roth make this volume available to the English literature. My role has been to edit extensively an English translation done in Prague by Margaret Sehierlova and to make some additions and modifications in several chapters not directly involving his patient series. Dr. Roth had kindly had me design these five chapters. Thanks are also given to Dr. Carlyle Smith of Trent University, Peterborough, Ontario, Canada, who proof-read the final typed manuscript. I am certain that Dr. Roth's monumental contribution to the literature will be received with appreciation which it deserves.

Roger Broughton, M. D.
Ottawa, June 1980

Preface

Pathological states of inappropriate or excessive sleep - narcolepsy and hypersomnia - are very frequent and cause the afflicted subjects considerable inconvenience and distress. The study of these hitherto inadequately elucidated conditions can help us to a better understanding of them and thus lead to more accurate diagnosis and to more effective treatment. The clinical, electroencephalographic and polygraphic investigation of these syndromes can also furnish valuable information concerning the physiology of both sleep and wakefulness.

When I began research on these conditions in 1949, I was guided by such considerations. Findings obtained in the initial 251 patients studied over a period of six years were summarized in a monograph entitled "Narcolepsy and Hypersomnia from the Aspect of the Physiology of Sleep", published in Czech in 1957, which contained extensive Russian and English summaries. A German translation was published in 1962. Although widely cited in the international literature, the full text of this book was unfortunately inaccessible to readers not knowing Czech or German. Consequently, I was frequently asked by many colleagues to publish an English translation. During the past 20 years, however.
there have been such scientific advances in the fields of
the physiology and the pathology of sleep that it has proven
necessary to rewrite the book in its entirety. I hesitated for a
long time before deciding to do so, as I was well aware of the
many difficulties involved. But I believe that the clinical experience
acquired and the findings obtained during 26 years of systematic
study of these problems could be of value to the medical
and scientific public and so decided to proceed. The present volu-
me is based on a study of 620 patients suffering from either
narcolepsy or hypersomnia that I have personally examined and
observed over many years at the Charles University Neurology
Clinic in Prague between 1949 and 1975. Many patients have been
studied for 15 to 20 years, and some for over 25 years, all with
careful follow-up. This series, I believe, is the largest of its kind
in the literature.

The purpO'se of the book is to present detailed information on
these two conditions which should be of use to physicians called
upon to treat them, i. e., neurologists, psychiatrists, general practitioners
and specialists in other fields. In this regard I have included
chapters on the neurophysiology, electroencephalography
and polygraphy of sleep, whic11 should also make the volume
easier for non-specialists to understand the main thrust of the
book.

Another purpose of t11e book is to provide specialists engaged
in sleep research with information acquiredin the study of my
material which, I hope, will be of so me value to them and perhaps
furnish new stimulation for clinical research. I quite realize that
some of my views and conclusions may turn out tO' be incorrect.
But I hope that, nevertheless, they will still prove of significance
for discussion. The next 20 years, I am convinced, will see such
scientific advances in this field that a completely new monograph
on the subject will once aga in be needed.

I should like to mention that the length of this book was fixed
beforehand and could not be exceeded. It was therefore necessary
to limit citations from the literature. I apologize to any authors
whom I have been unable to cite in full or at all for this reason.
Unfortunately, I also had to reduce the number and length of the
case histories, although many of them were so interesting that
they merited being reported in substantialy more detail.
With regard to the relative lengths of the various chapters, I have tried to be guided by their clinical and scientific importance. However, I have admittedly allotted more space to those aspects for which my findings and experience allow me to make a personal contribution.

Preface XV

I would like to take this opportunity to express my thanks to all those who helped me with this study. I am particularly indebted to my teacher, the late Professor K. Henner, former head of the Department of Neurology; and I am also very grateful to Professor J. Vymazal, M. D., D. Sc., the present head of the department, for his generous aid and support. Although I cannot name them all, I would like to thank my many colleagues for their helpful cooperation over the past years. Special thanks are due to Dr. S. Nevsimalov, C. Sc., who has worked with me now for many years, and to Dr. J. Faber, C. Sc. and Dr. Le van Thanh, C. Sc., for collaboration both stimulating and fruitful. Nor can I omit mentioning the many physicians who sent me their patients, the staff of the departmental EEG laboratory and all patients without whose voluntary participation our investigations would not have been possible.

Concerning the publication of the monograph, I would like to sincerely thank Mrs. Margaret Schierlova for her excellent work in the translation of the manuscript into English. I am deeply indebted to Professor Roger Broughton, M. D., Ph. D., of the University of Úttawa, Canada, for his invaluable help in editing this book. Without his assistance it would not have been published in English. He revised the text, made major suggestions on the physiological, neurochemical, therapeutic and other sections and gave much most helpful advice. He also wrote, together with me, chapters 1, 2, 4, 5 and 15 of this book.

And last, but by no means least, I wish to thank my wife Nacl'a for her boundless patience and understanding.

Prague, March 1980

Bedřich Roth