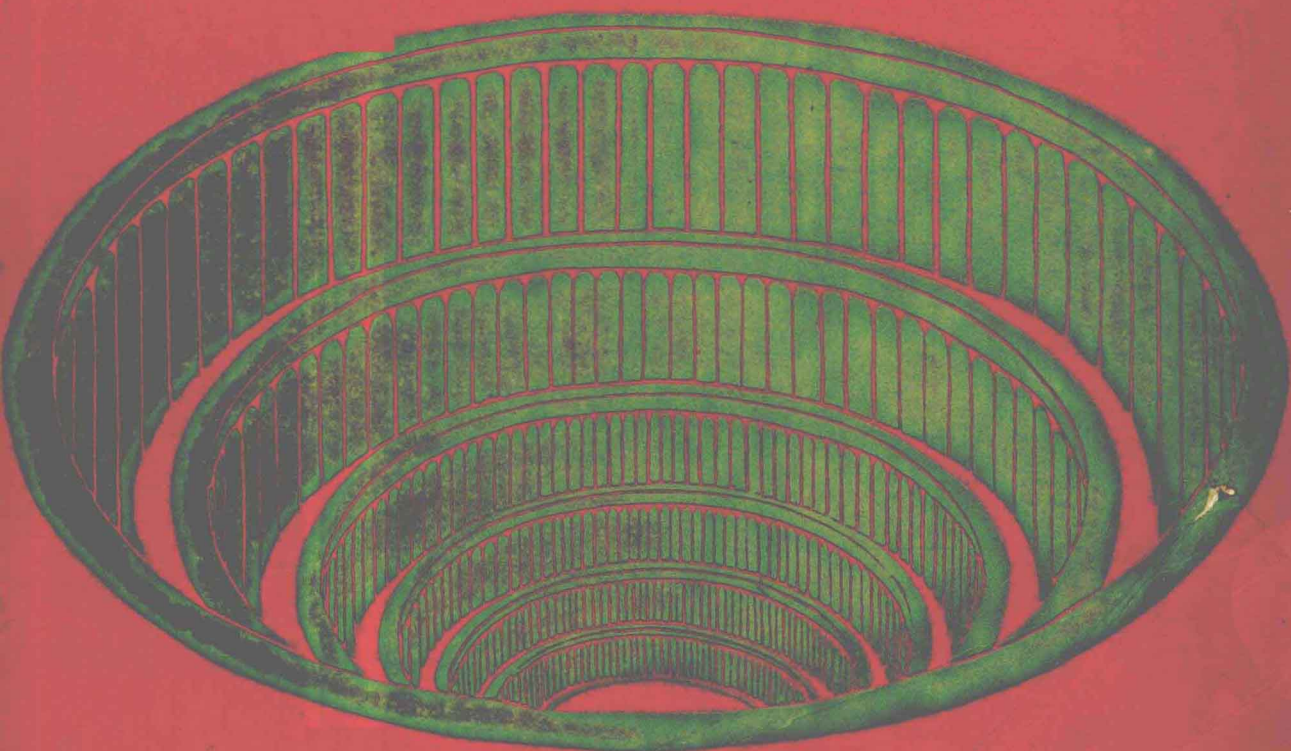


SYSTEMIC PATHOLOGY

SECOND EDITION

BY 38 AUTHORS

Edited by W. St C. Symmers



VOLUME 6
CHURCHILL LIVINGSTONE

Systemic Pathology

SECOND EDITION

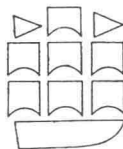
by THIRTY-EIGHT AUTHORS

VOLUME 6

Skin

Eyes

Ears



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CHURCHILL LIVINGSTONE

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Preface to the Final Volume of this Edition

This volume completes the second edition of *Systemic Pathology*. The four volumes anticipated when the preface to the first volume of this edition was written, in 1976, have become six. The publishers and the editor hope that the presentation of the text and illustrations may be considered to have done justice to the knowledge, skill and experience of the authors. That the book has been accepted as a useful addition to the literature of pathology is a realization of the intention of Professor George Payling Wright, of Guy's Hospital and the University of London, the senior editor of the first edition, who died in April, 1964. Payling Wright's death deprived the first edition, published at the end of 1966, of that uniformity of editorial scholarship that would have marked it throughout with the academic excellence that was the foundation of his own achievements as a teacher, research worker, philosopher and practitioner of pathology. It is not to be doubted that, had he lived to devote part of his retirement from active university life to the development of *Systemic Pathology*, the second edition would have been quite different in character from the work that has now appeared. Nevertheless, it may be hoped that this edition can be regarded as in some measure a tribute and a memorial to him.

* * *

In addition to the authors, many individuals have contributed to the preparation and production of this edition. Some are named in the chapters—many others are not, yet have done as much to make the book what it is. Among the unnamed are the members of the staff of Churchill Livingstone who were involved in preparing the book for publication. It is the publishers' policy that the work of their staff shall not be acknowledged other than in general terms: for this reason I may not thank these always helpful associates and friends by name but only through the expression of my appreciation of all that the publishers have put into the preparation of the six volumes. Anyone who is familiar with what is entailed in publishing, particularly medical publishing, which carries its peculiar responsibilities, will have some understanding of the complexity of the work entailed in developing this edition from the initial concept to the finished volumes: only those who have experienced it can appreciate fully the significance of the collaboration of publishers and authors, without which a work such as this could not be completed with any measure of success and pleasure.

As editor, I would thank not only the authors and the publishers, the blockmakers and the printers, but also those colleagues and friends who, most of them not directly involved in the book, allowed me the time to undertake what was quickly, and quite unforeseen, to become virtually a whole-time occupation. I am indebted above all to the staff of the Department of Histopathology of Charing Cross Hospital Medical School, London, and particularly to the present and past consultants—Dr J. Graham Jackson, Dr Bernard Fox, Dr Fernando J. Paradinas, Dr Brendan G. McCann and Dr Sami Shousha—who added my departmental duties to their own so that I might attend to the book. Successive Deans of Charing Cross Hospital Medical School—Dr Seymour J. R. Reynolds and Professor T. W. Glenister—and the Academic Board and the Council of the School tolerated a protracted elusiveness on my part that for long was accompanied by no material evidence that my time was being spent productively.

The invaluable and always immediate help of Mrs L. Shane Godbolt, Librarian to Charing Cross Hospital Medical School, and of other members of the staff of the Library, and of my technical and secretarial associates in the Department of Histopathology has been mentioned in the preface to Volume 1 or elsewhere in the book. I have been so dependent on their support that I would restate my debt to them here, although the words are an inadequate means of expressing how much I have appreciated the help and interest so freely provided.

Mr R. S. Barnett's photography has been exemplary. Without it each volume of the book would have

been appreciably diminished. His understanding of what is to be illustrated by a photomicrograph and his skill in depicting in a black and white photograph the diagnostic content of the original have more than complemented the text.

* * *

I have to note, with sadness, the death, since the preface to Volume I was written, of two associates of long standing who helped in the preparation of both editions of this book, Mr F. D. Humberstone and Mr H. Oakley. Mr Humberstone, first as head of the technical services of the Department of Histopathology of Charing Cross Hospital Medical School and later, when his physical health required a less demanding commitment, as head of the preparation laboratories in the School's Pathology Museum, contributed quietly, efficiently and loyally to the work of the School and Hospital during more than twenty years. Mr Oakley, at the time of his death, had been longer on the staff of Charing Cross Hospital Medical School than any other member: his kindness to the relatives of those who died in the Hospital, his skill as a mortuary technician, his knowledge of morbid anatomy and his loyalty to the Department of Histopathology are now part of the tradition of the School and of the Hospital.

* * *

Many colleagues—some of them friends or acquaintances, but most of them strangers—have taken the trouble to write or say that they welcomed the first volumes of this edition. A few have gone farther and pointed out improvements that they would have liked to see: such comments are helpful even if the suggestions that they contain cannot be adopted at the time or have to be turned down. Published reviews of this edition have been few, up to now, and most of them have been short and uncritical, if usually kind. It is a pity that the thoughtful review as a medium of constructive criticism has lapsed to the extent that it has: criticism based on a reviewer's experience and knowledge is valuable to author, editor and publisher—it is also welcome, and it should be encouraged.

* * *

There has been a surprisingly large number of requests for complimentary copies of particular chapters and even of entire volumes or of the whole book. It is proper to note, therefore, that it was not practicable to supply the authors with offprints of their chapters and that neither they nor the publishers, nor I, have copies available for free distribution.

* * *

It is not always realized that the costs of involvement in the preparation of a large textbook can be high. Few who write medical books expect to add appreciably to their income by this endeavour, though a considerable proportion will in fact find a small monetary profit. From discussion with several of the authors of *Systemic Pathology* it has been evident that little of their royalty would be left to them to share with the income tax collector after payment for secretarial help, bibliographical services, photography and postage. It does not seem right that the authors of a textbook like *Systemic Pathology* should be out of pocket by reason of their authorship: their collaboration is the more welcome for having been given uncomplainingly and without expectation of any significant return for their work other than its publication.

* * *

Some of those who have found the book of value have asked that acknowledgement should again be made of the support of one of their number without whose faith in the worth of the project this edition could have been neither initiated nor completed. Like them, I am grateful that such support, anonymous as it was, was also so timely.

* * *

In recording the completion now of this edition it is necessary to note the death during its preparation of two of its authors, William Henry McMeneney, who died on the 24th November, 1977, at the age of 72, and John Alexander Milne, who died on the 21st July, 1977, at the age of 57. Emeritus Professor McMenemey wrote the chapter on the pathology of the central nervous system for the first edition of *Systemic Pathology* and revised it, in collaboration with his colleague and former pupil, Professor W. Thomas Smith, for this edition. Professor Milne joined the team of authors of *Systemic Pathology* while the second edition was being planned: he intended to revise the chapter on the pathology of the skin that had been written for the first edition partly by Dr Henry Haber and partly, after Dr Haber's death in 1962, by the editor: as it turned out, the responsibility for completing this chapter became again the editor's obligation. McMenemey and Milne were pathologists in a great international tradition, each dedicated to the thoughtful practice of his specialty and to the advancement of knowledge in the field that he had made his own: their contributions to this book are among the final statements of their experience and authority.

* * *

Sir Roy Cameron, shortly before his death in October, 1966, wrote in his foreword to the first edition of *Systemic Pathology*, 'These volumes have an immediate and universal usefulness throughout the world of Medicine'. I hope it may not appear presumptuous to echo these words now that the second edition is completed. If those who make use of these six volumes find Cameron's opinion of the original edition to apply to its successor, perhaps they will remember that the knowledge in these pages is founded on the work of such scholars as Cameron himself and his great predecessors and contemporaries 'throughout the world of Medicine'.

WILLIAM ST CLAIR SYMMERS

Northwood
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January, 1980

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§

Acknowledgements for Illustrations

The caption of each illustration that requires an acknowledgement includes the symbol §, printed after the figure number (for example, Fig. 40.10.§). A footnote on the page on which the first such illustration in each chapter appears refers the reader to the page on which the acknowledgements are made. In the lists of acknowledgements, all the illustrations from each source are grouped together, in numerical sequence.

Some of the authors of the book and some of those who provided them with illustrations for the first volumes of this edition have found this method of acknowledgement to be inadequate. The § footnote, appearing only once in each chapter, is liable to be overlooked: this has led to a mistaken impression that acknowledgements have not been made in all instances requiring them.

The decision not to include acknowledgements in the captions of the illustrations but to collect them at the end of the chapter was made by the editor, who is responsible for the embarrassment caused to the authors and to those who helped to illustrate the book.

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39: *The Skin*

by H. HABER and W. ST C. SYMMERS

revised by J. A. MILNE and W. ST C. SYMMERS

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39: *The Skin*

by H. HABER* and W. ST C. SYMMERS

revised by J. A. MILNE† and W. ST C. SYMMERS

Note: Through Professor Milne's death, the final revision of this chapter had to be completed without his detailed consideration of each statement, which, had he lived, would have ensured that the text was completely in conformity with the views that reflected both his long experience as a practising specialist in the pathology of the skin and his encyclopaedic knowledge. It is therefore proper to note that views expressed in the chapter as it now appears are not necessarily views that Professor Milne would have put forward or accepted.

W. St C. S.

INTRODUCTION

The skin envelops the body and forms the first line of defence against the environment. It is equipped with a complicated system of nerve-endings that communicate information concerning our surroundings, and in this way it helps to protect the body from harm. Its blood vessels and sweat glands are of great importance in the control of body temperature. The sebaceous glands elaborate secretions that flow into the hair follicles and thence reach the skin surface; the sebum formed by these glands helps to protect the skin from dessication and is also thought to possess bactericidal and fungicidal powers.¹

Because of its exposed position, the skin is subject to many different types of injury—physical, chemical, ectoparasitic and infective in origin. These various insults are the origin of many of the dermatoses.

Apart from the exogenous causes of skin disease, disorders of any system in the body may manifest themselves in the skin. Indeed, changes in the skin may be the first clinical evidence of certain diseases of other parts of the body. It is significant too that the cutaneous manifestations of some important general diseases were described before other tissues and systems were recognized to be involved. For example, lupus erythematosus is now known to be a systemic disease of the connective tissues,² sarcoidosis a systemic granulomatosis³ and urticaria

pigmentosa a manifestation of a systematized mast-cell proliferation.⁴ Angiokeratoma corporis, which until quite recently was a dermatological curiosity, is now known to be a serious inherited disorder of lipid metabolism mediated by the X-chromosome as a recessive characteristic.⁵⁻⁷

By virtue of its blood vessels and sweat glands, the skin is able to reveal emotional states, as in the pallor of fright or anger and the flush of anger or embarrassment. Sweating is also sometimes an effect of emotional changes. Both of these rapidly labile features of the skin—blood flow and perspiration—are related to its autonomic innervation, for the autonomic system controls the cutaneous blood vessels and the sweat glands.

Because the skin is so readily accessible to inspection it is hardly surprising that dermatology became the richest field of descriptive clinical medicine. The very range and diversity of the many different appearances presented by pathological changes in the skin have commonly, and perhaps unavoidably, led dermatologists to label them with long names that can be a source of confusion to colleagues in other specialties.

The histopathology of the skin occupies an important place in general pathology and immunology. Because of its specialized structure and functions, the skin presents numerous types of lesions. Many of the diseases of the epidermis, such as dermatitis, psoriasis and lichen planus, have features without parallels in other fields of human pathology. In contrast, the diseases of other body systems that also involve the skin can be studied

* Henry Haber: 3rd October 1900–7th July 1962.

† John Alexander Milne: 19th February 1920–21st July 1977.

with advantage, both clinically and histologically, in their dermatological manifestations because the latter are readily visible and accessible. It is a pity that greater use has not been made of this easily available material, for the skin is one of the few organs that serial biopsies can be safely obtained from, thus enabling the progress of disease to be followed. This is particularly true of lymphomas

(for instance, as exemplified in the natural history of mycosis fungoides and related conditions) and of the immune disorders of connective tissue.

It cannot be stressed too strongly that clinical dermatology and the histopathology of the skin are inseparable facets of the same subject: knowledge of the former is indispensable for an understanding of the latter, and *vice versa*.

THE STRUCTURE OF NORMAL SKIN

The skin is developed from both ectoderm and mesoderm. The ectoderm is the origin of the epidermis, the skin appendages, and the neural elements. The mesoderm is the origin of the dermis—vessels, connective tissue and muscle.

The Epidermis

The epidermis—the superficial cellular portion of the skin—consists of two types of cell, the keratinocytes and the dendritic cells. These are considered in some detail below. The dendritic cells (see page 2530) include the melanocytes, the Langerhans dendritic cells and cells at present known as the indeterminate dendritic cells. The keratinocytes, which hitherto have commonly been referred to simply as the ‘epidermal cells’, are disposed in four named strata—the basal cell layer, the prickle cell layer, the granular layer and the cornified layer (see below). In most of the skin the thickest of these strata is the prickle cell layer. This is so named because its cells, when examined by the light microscope, appear to be joined together by intercellular bridges, sometimes termed prickles. This appearance of bridging gives a false impression that the epidermis is a syncytium (Fig. 39.1). Electron microscopy has shown that in fact there is no physical continuity between the cells and that the so-called intercellular bridges are simply points of contact between adjacent epidermal cells. Where the epidermal cells make contact with each other there is a localized thickening of the cell membrane that is known as the desmosome, into which the intracellular tonofibrils are inserted.⁸ Between adjoining desmosomes there is a layer of apparently structureless material, the cement substance. The cells of the basal layer of the epidermis have half desmosomes where they make contact with the dermis. This form of attachment of the epidermal cells to each other gives the epidermis great mechanical strength and allows it to function as a continuous sheet. Since the

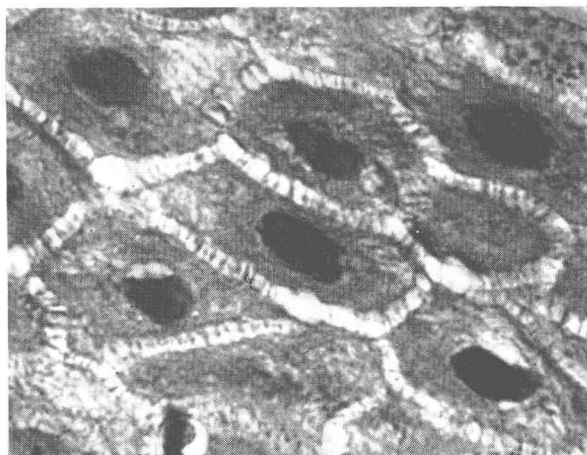


Fig. 39.1. § Normal skin. Cells of the prickle cell layer, showing the intercellular prickles. *Haematoxylin-eosin*. $\times 2000$.

epidermis is avascular its metabolic activity is maintained by diffusion of plasma from the blood vessels of the dermis through the tissue spaces between the desmosomes.

One of the important functions of the epidermis is the production of keratin. This fibrous protein forms the outermost covering of the body, and by virtue of its physical characteristics it protects the underlying skin against damage by physical and chemical agents. Keratin is formed continuously by the epidermal cells. Although the ultimate molecular structure of keratin may be the same in all situations, its mode of formation and its microscopical appearance differ in different areas. Thus, the keratin of the palms and soles is different from that of, for instance, the back of the trunk (see next page). Again, the keratin over the extensor aspect of the knees and elbows is different from that of the flexures.⁹

The thickness of the epidermis varies greatly from site to site. It is greatest on the palms and soles and thinnest on the glans penis and the eyelids.

§ See *Acknowledgements*, page 2820.