Second Edition VOLUME I

DIAGNOSIS OF DISEASES OF THE CHEST

FRASER

PARÉ

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PREFACE TO THE SECOND EDITION

It was stated in the Preface to the First Edition that the book was written with the aim of defining an approach to the diagnosis of diseases of the chest based on the abnormal roentgenogram and of emphasizing the value of the roentgenogram as the *first* rather than the *major* step in diagnosis. Our experience during the years since publication in 1970 has not caused us to alter these principles. However, we wish to reiterate that the emphasis we place on the roentgenogram as the first step in reaching a diagnosis does not represent an attempt to relegate history and physical examination to a position of no importance, but merely to place them in proper perspective. We have never ceased to recognize that an intelligent integration of information from roentgenologic, clinical, laboratory, and pathologic sources is essential to the diagnosis of chest disease, and we trust that the approach we have taken in this second edition clearly illustrates that conviction.

Because of the vast amount of new knowledge that has accumulated since the publication of the first edition and of a number of important omissions from the first edition, simple revision has proved impossible. It has been necessary to rewrite the book almost completely. Inevitably, its length has increased and the original two volumes have expanded to four. We have regretted this because we recognize the danger of the book's becoming purely a reference work rather than a textbook. However, we have continued to hope that the organization of the book will enable readers to be highly selective in the material they wish to read. The subdivision of virtually all descriptions of chest disease into etiology, pathogenesis, pathologic characteristics, roentgenographic manifestations, and clinical manifestations should permit readers to cull the material appropriate to their disciplines. For example, a substantial part of the research in chest disease in recent years has been concerned with pathophysiology, and we felt obliged to include much of this highly pertinent information in the text. Since such material is indicated by appropriate headings, readers can choose whether they wish to delve deeply into this aspect of any disease.

The first three volumes contain the 18 major chapters of the book. Volume I includes descriptions of the normal chest, methods and techniques of investigation, clinical features and roentgenologic signs of chest diseases in general, and diseases of developmental origin. Volume II deals with the infectious diseases, immunologic disorders, neoplasms, thromboembolic disease, and pulmonary hypertension and edema. In Volume III, we have grouped together environmental and airway diseases, abnormalities caused by external physical agents, diseases of unknown origin, and abnormalities of the pleura, medi-

astinum, chest wall, and diaphragm. The final chapter deals with respiratory disease associated with a normal chest roentgenogram. The tables of differential diagnosis, formerly in Volume I, now constitute a major portion of the smaller Volume IV. This volume also contains a number of tables of normal values in roentgenologic, biochemical, and function test assessment, and other material that we thought should be available for ready reference.

A new approach to the differential diagnosis of chest disease has been developed in the form of "decision trees," also included in Volume IV in association with the tables of differential diagnosis. Since publication of the first edition, it has become apparent that use of the tables of differential diagnosis has been largely restricted to radiologists, chiefly those in training. As a means of involving the chest physician in this exercise, we have designed "decision trees," which incorporate into the equation the clinical presentation of the patient. As in the tables of differential diagnosis, the patterns of disease are subdivided into a number of diagnostic possibilities, each of which follows a different "branch" of the tree, and the most likely diagnoses are color-coded. The presenting symptoms and signs of each disease likely to produce the specific roentgenographic pattern are indicated at the base of each branch, followed up the branch by the appropriate diagnosis, and at the top by the major methods of confirming the diagnosis. We hope that this method of presentation will find favor with our more clinically oriented readers.

In addition to updating virtually all sections of the book, we have made major additions and revisions in a number of specific areas. There are detailed descriptions of the geometry, dimensions, and morphology of the conductive, transitory, and respiratory zones of the lung and the vascular system; blood gases and acid-base balance; the nonrespiratory functions of the lung; development of the lung; diagnostic applications of ultrasonography; mass chest roentgenography as a screening procedure; the techniques, indications, yield, and complications of lung biopsy; new methods of evaluation of pulmonary function, particularly of the small airways; host defense mechanisms in bronchopulmonary infections, including a thorough discussion of the compromised host, opportunistic infections, and host-etiology relationships; prognosis in neoplastic disease of the lungs; and drug-induced pulmonary disease. In addition, there is a complete reorganization of the chapter on pulmonary abnormalities of developmental origin, a complete revision of the section on pulmonary edema, a revision of the chapter on occupational diseases to conform to the UICC-ILO classification, and extensive additions to the discussion of the obstructive airway diseases, particularly with regard to their pathophysiology.

The burgeoning of knowledge in the field of chest disease since 1970 has been astonishing. The 4,300 odd references that made up the bibliography in the first edition were selected from 19 journals and several books that had been published during the previous 15 years. In the 7 years since publication of the first edition, more material on chest disease appeared in the same journals and in a number of new books than had been written in the previous 15 years. We thus faced the rather prodigious task of reviewing several thousand new articles and of selecting those whose content we felt was appropriate for inclusion in the manuscript and in the bibliography as sources for further reading. As a result, the bibliography has more than doubled since the first edition. We suspect that we could be justly criticized for not being more selective in our choice of referenced material, but we wish to emphasize that the articles cited, although numerous, represent a relatively small proportion of the total output. Each of the first three volumes in the second edition will

have its own list of references. Since new references were added to the manuscript late in the writing, such material has of necessity been cited with out of sequence numbers. However, all references are numbered consecutively if not sequentially.

At the end of the Preface to the First Edition, we invited our readers to inform us of differences of opinion they may have had with the contents of the book. Many were kind enough to write us, pointing out several errors and suggesting ways and means of improving the book generally. Several of these suggestions have been effected in this second edition. Again, we invite our readers to express their opinions and offer their advice. To reiterate a statement made in the original Preface, it is only through such interchange of information and opinion that we can hope to establish on a firm basis the knowledge necessary for a full understanding of respiratory disease.

R. G. F.

J. A. P. P.

ACKNOWLEDGMENTS

Although it might be assumed that the preparation of the second edition of a book is a relatively simple task compared with preparing the first, our experience has proved that assumption erroneous. The 4 years required for the production of the first edition expanded to at least 5 for the second, providing clear evidence that even with experience the complexities involved in the production of a book of this magnitude are difficult to appreciate. The writing of manuscript and the choice and preparation of new illustrations were the most formidable part of the undertaking, but the many steps necessary to the final product required the unselfish and enthusiastic contributions of many hands and minds, and the support and encouragement we received from many of our friends are greatly appreciated and duly acknowledged.

It is not possible to overstate our gratitude to our secretaries, who handled magnificently the tedious and necessarily exacting task of transcribing manuscript from tape, typed the several drafts up to and including the final manuscript, and cheerfully coped with all the innumerable problems encountered. Mrs. Joan Bell, Miss Bridget Byrne, and Mrs. Jean Farrel of the Royal Victoria Hospital in Montreal and Ms. Carolyn Lehman and Miss Sheila Walker of the University of Alabama Medical Center in Birmingham all exhibited exemplary patience and devotion in accomplishing this thorny chore. With some help, these assistants also carried out the tedious job of recording, filing, checking, and final validation of the more than 5,000 new references, an extremely frustrating chore that they performed with meticulous accuracy. The devotion and diligence with which they carried out their various tasks is deeply appreciated. We are also grateful to Ms. Ursula Matthews for the skill with which she subedited several chapters of the book.

Many of our medical colleagues were kind enough to review sections of the manuscript and to offer advice for their improvement. Special thanks are due to Drs. Peter Macklem, Stefan Vas, Len Moroz, John Seely, Rudy Dollfuss, James Hogg, Peter Paré, Jr., and Fred Winsberg for their valued counsel and discerning criticism.

The majority of case histories and roentgenograms reproduced here are of patients of members of the Attending Staff of the Royal Victoria Hospital. Our indebtedness to these friends and colleagues cannot be overemphasized, not only for their generosity in permitting us to publish these case reports but also for the benefit of their experience and guidance over the years.

During the period of writing, it was inevitable that the time expended by the authors on teaching and clinical responsibilities was reduced significantly. These additional responsibilities were added to the already overburdened shoulders of our colleagues in the Departments of Diagnostic Radiology and Medicine of the Royal Victoria Hospital and Queen Mary Veterans Hospital of Montreal, and the Department of Radiology, the Univer-

sity of Alabama Medical Center in Birmingham. Their contributions to the book, although indirect, are sincerely appreciated.

The superb photographic work throughout these volumes was the accomplishment of Ms. Pat McKenna and her staff, particularly Ms. Edith Boltz, of the Department of Visual Aids of the Royal Victoria Hospital and Mr. Charles Walton of the Department of Radiology, University of Alabama Medical Center in Birmingham. Their craftsmanship and rich experience in photography are readily apparent in these pages. Most of the graphs and diagrams were charted with meticulous accuracy by Mr. Lionel Bartlett; the majority of function tests were performed with care and devotion by Mr. J. Nowaczek.

We are indebted to Mr. Douglas McDonald, Executive Director of the Royal Victoria Hospital, for arranging financial assistance toward the cost of illustrations. Throughout our labors, we have received tremendous support and cooperation from the publishers, notably Mr. Jack Hanley, Ms. Kathy Pitcoff, and Ms. Evelyn Weiman, who effectively and sympathetically minimized the many obstacles we encountered.

Finally, and with immense gratitude, we recall the patience and understanding displayed by our wives and children throughout our labors. Without their continuous encouragement, this book surely would not have been completed, and we acknowledge their many virtues with much love.

R. G. F.

J. A. P. P.

PREFACE TO THE FIRST EDITION

This book was written with the aim of defining an approach to the diagnosis of diseases of the chest based on the abnormal roentgenogram. Experience over the years has led the authors to the conclusion that the chest roentgenogram represents the focal point or sheet anchor in the diagnosis of the majority of pulmonary diseases, many patients presenting with either no symptoms and signs or entirely nonspecific ones. This emphasis on the roentgenogram as the first step in reaching a diagnosis does not represent an attempt to relegate history and physical examination to a position of no importance, but merely an effort to place them in proper perspective. In no other medical field is diagnosis so dependent upon the intelligent integration of information from roentgenologic, clinical, laboratory, and pathologic sources as in diseases of the chest. We submit that the roentgenogram is the starting point in this investigation; the knowledge of structural change thus obtained, when integrated with pertinent clinical findings and results of pulmonary function tests and other ancillary diagnostic procedures, enables one to arrive at a confident diagnosis. Some patients manifest symptoms and signs that themselves are virtually diagnostic of some chest disorders, but even in such cases the confirmation of diagnosis requires the presence of an appropriate roentgenographic pattern.

A glance through the pages will reveal an abundance of roentgenographic illustrations that might create the illusion that this book is written primarily for the roentgenologist, but this is not our intention. In fact, the clinical, morphologic, and laboratory aspects of many diseases are described at greater length than the roentgenologic, a fact pointing up the broad interest we hope the book will engender among internists, surgeons, and family practitioners interested in chest disease. The numerous illustrations reflect the aim of the book—to emphasize the value of the roentgenogram as the *first* rather than the *major* step in diagnosis.

During the writing of the book, our original plan was considerably modified as the format unfolded and we became even more aware of the complexities of design and organization. Originally, our approach to differential diagnosis suggested a division of chapters on the basis of specific roentgenographic patterns. It soon became apparent, however, that since many diseases give rise to various different roentgenographic patterns, this method of presentation would require tedious repetition of clinical and laboratory details in several chapters. To obviate this, we planned tables of differential diagnosis, listing etiologic classifications of diseases that produce specific roentgenographic patterns and describing briefly the clinical and laboratory characteristics of each disease, thus facilitating recognition of disease states.

The tables are designed to be used with the text in the following manner. When a specific pattern of disease is recognized, the appropriate table should be scanned and those conditions selected that correspond most closely with the clinical picture presented by the patient. Additional information about the likeliest diagnostic possibilities can be obtained by referring to the detailed discussions in the relevant sections of the text (page numbers are cited after each diagnosis). The tables relate to 17 basic patterns of bronchopulmonary, pleural, and mediastinal disease; they are grouped together in Chapter 5 in Volume I and may be located with ease from the black marks found on the upper corners of their pages. Each table is preceded by a detailed description and representative illustrations of the specific roentgenographic pattern. An attempt has been made to indicate the relative incidence of the diseases.

Although our original plan called for a one volume presentation, it soon became apparent that the length of the text and the number and size of illustrations necessary for full coverage of the subject required two volumes. Volume I includes descriptions of the normal chest, methods and techniques of investigation, clinical features, and roentgenologic signs of chest diseases, the tables of differential diagnosis, and chapters devoted to diseases of developmental origin and the infectious diseases; in Volume II appear detailed discussions of the morphologic, roentgenologic, and clinical aspects of all other diseases of the thorax arranged in chapters according to etiology.

The roentgenograms have been reproduced by two different techniques, the majority in Volume I by the logEtronic method and those in Volume II by direct photography. The publishers have been generous in allotting sufficient space for the reproduction of the roentgenograms in a size adequate for good detail recognition.

Much of the material in the book has been based on our personal experience gained in the past almost two decades, during which we have had a predominant interest in pulmonary disease. Obviously, this experience has been greatly enhanced by the extensive literature that has accumulated during these years, and we are mindful of the tremendous help we have received from the contributions of others. Our free use of the literature is reflected in the extensive bibliography.

Certain differences from the contents of other books on respiratory disease will be noted. First, this text contains no reference to treatment. Since drug therapies and surgical techniques are constantly changing, any attempt to include them would make the book out of date almost before it was published. Second, we have intentionally made only passing reference to pulmonary disease peculiar to children, a full description of which would require a complete separate text.

The relative incidence of respiratory diseases has changed considerably over the last quarter century. In some diseases, such as tuberculosis and bronchiectasis, a decreased frequency reflects improved public health measures and therapeutic innovations; in others, man's therapeutic triumphs have proved a mixed blessing, enabling patients with disabling chronic respiratory disease to live longer despite formerly fatal pneumonias. Perhaps even more important, man himself is responsible for varying the spectrum of respiratory disease as a result of his irresponsible insistence upon increasing the amount and variety of atmospheric pollutants. Inhaled contaminated air not only is regarded as the major etiologic factor in chronic obstructive pulmonary disease and the inorganic dust pneumoconioses, but also has been incriminated in the etiology of several hypersensitivity diseases of the lungs.

This last group comprises the "extrinsic" form of allergic alveolitis. The number of conditions involved, when added to the better known "intrinsic" counterpart—the collagen diseases—is largely responsible for the length of the chapter devoted to immunologic diseases. Other changes that have contributed to the "new face" of pulmonary disease include increasing knowledge of the hormonal effects of neoplasms; the discovery that various immunologic defects may reduce host resistance to infection; and finally the appearance in the western world of parasitic infestations and bacterial infections formerly considered so rare in those areas as to warrant little consideration in differential diagnosis, but now of some importance because of the modern day ease of intercontinental travel. Although the novelty of these recent changes may have led the authors to consider them in greater detail and length than is their due, the emphasis may serve to bring them into proper perspective.

Finally, we recognize our fallibility. It is inevitable that some observations in a text of this magnitude will prove erroneous in time or will find disagreement among our knowledgeable readers. This we expect and accept. We sincerely hope that such differences of opinion will be made known to us, so that they may be weighed and, where appropriate, introduced into subsequent editions or revisions. It is only through such interchange of information and opinion that we can hope to establish on a firm basis the knowledge necessary to a full understanding of respiratory disease.

R. G. F.
J. A. P. P.

CONTENTS

VOLUME I

Chapter 1

THE NORMAL CHEST

INTRODUCTION

Description of the anatomy of the normal chest as viewed roentgenographically usually are concerned with gross morphology in relation to roentgenology, with emphasis on such details as the spatial distribution of the bronchial and arterial trees, the location of bronchopulmonary segments, and the configuration of the diaphragm. Although such information is essential to a thorough understanding of roentgen pathology, it fails to take into consideration the minute structure of the lung, which is generally relegated to the province of the histopathologist. This lack of emphasis on peripheral anatomy ignores the fact that the majority of pulmonary diseases that alter roentgenographic density involve the lung parenchyma; and that understanding them therefore requires knowledge of peripheral lung structure and the roentgenographic appearance of involvement of individual parenchymal units.

Since the prime purpose of this book is to describe roentgenographic patterns and to discuss their differential diagnosis, little purpose would be served by reviewing in great detail the physiology of respira-

tion, which has been done so well by others.[1-3] Nevertheless, a truly proficient interpretation of the roentgenogram requires not only familiarity with the anatomy of the lung but also a knowledge of pulmonary physiology, a requirement that is even more necessary in view of the ever-increasing number and complexity of dynamic roentgenographic procedures.

With these points in mind, this chapter begins with a concise account of the minute structure of the lung, dealing in turn with its morphology, roentgenology, and function.

THE AIRWAYS AND PULMONARY VENTILATION

General Considerations

The function of the bronchial tree is to conduct air to the alveolar surface, where gas transfer takes place between respired air and gas dissolved in the blood of the alveolar capillaries. The inspired air should be evenly distributed to the alveolar capillary bed, with minimal resistance to flow. The greater part of the length and the smaller part of the volume of the respiratory tract are concerned only with the conduction of air; this part comprises the trachea and bronchi (whose walls contain cartilage) and nonalveolated bronchioles (no cartilage). The remainder of the respiratory tract, comprising the large bulk of the lungs, is concerned with both conduction and gas exchange, the terminal unit (the alveolus) being the only structure whose function is uniquely that of gas exchange. Thus the airway system can be subdivided into three zones, each with different structural and functional characteristics.

The conductive zone includes the trachea, bronchi, and nonalveolated bronchioles in which air cannot diffuse through the well-developed wall. The nonparenchymatous pulmonary structures constitute this zone along with the pulmonary arteries and veins, lymphatic channels, nerves, connective tissues of the peribronchial and perivascular spaces, the interlobular septa, and the pleura.

The transitory zone, as its name implies, carries out both conductive and respiratory functions. It consists of the respiratory bronchioles, alveolar ducts, and alveolar sacs, all of which conduct air to the most peripheral alveoli. In addition, alveoli that arise from their walls are the site of gas exchange. This zone and the respiratory zone constitute the parenchyma, the spongy respiratory portion of the lung.

The respiratory zone consists of the alveoli, whose sole function is the exchange of gases between air and blood.

The descriptions that follow of the geometry, dimensions, and morphology of these zones are based on material gleaned largely from two sources—the publications of Horsfield, Cumming, and their associates;[4-6] and the 1963 monograph, "Morphometry of the Human Lung," by E. R. Weibel.[7] The morphometric data were derived by Weibel from studies of five normal human lungs obtained at necropsy from subjects whose ages ranged from 8 to 74 years; the conductive elements were studied macroscopically from plastic casts of the human bronchial tree fixed at approximately three-quarters total lung capacity (TLC); and the respiratory and transition zone structures were examined microscopically from lungs fixed with concentrated formalin steam at about the same volume. The data recorded by Horsfield, Cumming, and colleagues were from a pair of normal lungs removed from a 25-year-old man. The lungs were inflated and fixed at a volume of 5 liters corresponding to a position of inspiration, and the airways (complete down to the terminal bronchiolar level) were cast in nonshrinking resin.

THE CONDUCTIVE ZONE

Geometry

Cumming and his associates[5] point out that usually, in nature, the distribution and collection of nutrients requires a branching system, and that biological branching systems show great similarity. For example, deciduous trees must provide large surface areas of leaves exposed to sunlight, so that photosynthesis can proceed with appropriate supplies transported from the ground. Observing a large oak in winter and in summer reveals how well the tree's branching system accomplishes this. Similar circumstances exist in the lung, in which a branching system of conducting airways is essential to transport the max-

imal amount of air, with minimal resistance, to the respiratory zone. Among the methods for comparative description of biological branching systems, the ones most conveniently applicable to the lung probably are those proposed by Strahler[8] and by Horsfield and Cumming.[4]

The basic branching pattern of the bronchial tree is dichotomous (*i.e.*, the parent branch divides into two parts). In any such system, the branching may be symmetric (two branches equal in all respects) or asymmetric (variation in the diameter or length of branches in a given generation, or in the number of divisions to the end branches, or a combination of these). In the bronchial tree, there is variation in both branch diameters and the number of divisions; thus, the system is one of *asymmetric dichotomy*. If, in an asymmetric dichotomous branching system, one counts distally from the largest to the smallest branch along two pathways of different length, at some point one reaches a generation whose number is the same in both pathways but whose functional and morphologic characteristics are different. (For example, compare a pathway in a lower-lobe basal segment supplying parenchyma in the base of a lung with a "lateral pathway" arising from a large segmental branch supplying contiguous parenchyma in the inner third of the lung.) For this reason, Horsfield and Cumming[4] suggested that the traditional method of numbering generations of bronchial branches distally from the trachea is unsuitable for such an asymmetric system. Instead, they proposed counting generations proximally from branches of arbitrary but uniform diameter (0.7 mm in their study), a method that reveals a close relationship between the diameter of a branch, the number of distal respiratory bronchioles it supplies, and its generation number (Figure 1–1). Thus, with such a system one can describe accurately the dimensions of the tracheobronchial tree—the number of branches, and their length, diameter, and volume.

Both the Strahler and Horsfield-Cumming systems employ this method of counting proximally from the smallest branch. The first step is to identify the smallest branch—in a tree, the twig that bears the leaf, and in the pulmonary airways, the alveolar sac. This structure is considered to be of order 1. When two such structures join, they form a single branch (order 2); when two of order 2 join, they form an order 3, and so on. (For the sake of clarity, "generation" is applied to divisions counting distally from the trachea, and "order" to divisions counting proximally.) Difficulties arise, however, when branches of two orders join, and it is at this point that the systems differ. In the Strahler system (Figure 1–2) when different orders join, the larger number continues unchanged, thereby providing little information about the total number of branching points, whereas in the Horsfield-Cumming system (Figure 1–3) the larger order number increases by one. Thus, the segment proximal to the junction of a second-order and sixth-order branch would be sixth order according to the Strahler system and seventh order according to the Horsfield-Cumming system.

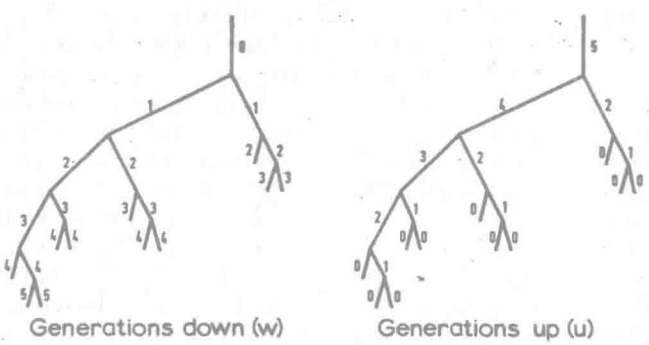

Figure 1–1. Generations in an Asymmetrical Dichotomously Branching System. Two methods of numbering are shown: the branches may be numbered downward starting with the mainstem as 0 (*on the left*), or, alternatively, may be numbered upward starting with the end branches as 0 (*on the right*). (Reprinted from Horsfield, K., and Cumming, G., J. Appl. Physiol., *24*:373, 1968, with permission of the authors, editor, and publisher.)