FRASER AND PARÉ

DIAGNOSIS OF DISEASES OF THE CHEST

An Integrated Study Based on the Abnormal Roentgenogram

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An Integrated Study Based on the Abnormal Roentgenogram

VOLUME I

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This book is dedicated
to the memory of

DR. FELIX G. FLEISCHNER

PIONEER, INSPIRED TEACHER,
DEDICATED PHYSICIAN AND RADIOLOGIST,
A HUMBLE AND COMPASSIONATE HUMAN BEING

PREFACE

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 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 that points 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 which 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 which 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 Five in Volume I and may be located with ease from the black marks which stain 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 during the past almost two decades 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, 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, and 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.

ACKNOWLEDGMENTS

It is difficult, even in retrospect, to appreciate the complexities involved in the production of a book of this magnitude. Although the writing of manuscript and the choice and preparation of illustrations proved the most formidable part of the undertaking, 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 who urged us forward in our task is greatly appreciated. To acknowledge all separately would make tedious reading, and therefore we say "Thank you" to everyone who helped us and restrict individual acknowledgment to those who have contributed time and effort.

It is not possible to overstate our gratitude to our secretaries, Miss Coni Reed and Miss Elizabeth McKee, who handled magnificently the tedious and necessarily precise task of transcribing manuscript from tape, typed the several drafts up to and including final manuscript, and cheerfully coped with all of the innumerable problems encountered. Their patience and devotion in accomplishing this thorny chore has been exemplary. During the final period their task was somewhat lightened by the able assistance of Mrs. Jean Riti and Miss Madeleine Miller, who also maintained the same cheerful acceptance of heavy workloads.

At the inception of the book, Miss Ursula Matthews accepted the awesome task of editing manuscript, and we are deeply grateful to her for seeing the job through to completion, despite a move to another city before the book was finished. She reduced the literary ineptness of the authors to a minimum without altering the meaning of the written word, and we are most grateful for her talent. The tedious job of recording, filing, checking, and final validation of the almost 4500 references was carried out with meticulous accuracy by Miss Catherine Mayhew and Mr. Richard Fraser, often in extremely frustrating circumstances. The devotion and diligence with which they carried out this task is deeply appreciated.

Dr. W. M. Thurlbeck deserves special mention for his invaluable contributions, specifically the descriptions of morphology in Chapters 1 and 2 and the legends for the majority of the pathology illustrations throughout the book. In addition, he critically reviewed Chapters 8 and 12, and his many helpful suggestions resulted in modification of these and other sections. His substantial contributions are most gratefully acknowledged. Several 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, John Henderson, and Margot Becklake 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. We wish to acknowledge particularly the assistance given by certain physicians who devoted time and effort to providing roentgenograms and case reports: Dr. George Genereux of the University of Saskatchewan, Saskatoon; Dr. David Berger and Dr. D. D. Munro of the Royal Edward Chest Hospital, Montreal; and Drs. J. F. Meakins, R. E. Donevan, and R. E. Dollfuss of the Royal Victoria Hospital.

During the period of writing, it was inevitable that the time expended by the authors on teaching and clinical responsibilities within the Departments of Diagnostic Radiology and Medicine was reduced significantly. These additional responsibilities were added to the already overburdened shoulders of our colleagues in these two Departments, who accepted them without objection. Their contributions to the book, although indirect, are sincerely appreciated.

The superb art work throughout these volumes was the accomplishment of J. Gebhardt Smith, medical artist of the Royal Victoria Hospital, whose craftsmanship and rich experience in medical illustration is readily apparent in these pages. We are indebted to her for the expertise with which she culled from our vague descriptions the true sense of what we wished to depict. The majority of the diagrams were charted by Mr. Lionel Bartlett, chief respiratory technician of the Royal Victoria Hospital, with his usual excellent draftsmanship.

We are indebted to Dr. J. Gilbert Turner, former Executive Director of the Royal Victoria Hospital, for arranging financial assistance, particularly toward the cost of illustrations. And, throughout, we have received tremendous support and cooperation from the publishers, notably Mr. John Dusseau, Mr. Michael Jackson, and Mr. Sam Mink, 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.

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VOLUME I

VOLUME II

1

THE NORMAL CHEST

INTRODUCTION

Descriptions 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 dia-

phragm. 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. We are of the opinion that this lack of emphasis on peripheral anatomy is erroneous: since the majority of pulmonary diseases that produce alteration in roentgenographic density involve the lung parenchyma, we feel that our attention should be directed first toward 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 respiration, 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 of dynamic roentgenographic procedures that are being performed.

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, its roentgenology, and its function.

THE LUNG UNIT

THE ACINUS AS AN ANATOMIC UNIT

In the bronchial pathway to the periphery of the lungs, the last purely conducting structure is the terminal bronchiole. The portion of the lung distal to the terminal bronchiole is the *acinus*, which comprises respiratory bronchioles, alveolar ducts, alveolar sacs, and alveoli (Figure 1–1). It is in this region that gas exchange takes place. The precise arrangement and exact number of structures that form the acinus are not known, since reconstruction is extremely difficult and tedious. However, it is known that respiratory bronchioles have some alveoli in their walls and that cuboidal epithelium lines the lumen between alveoli. Progressively more alveoli are present in succeeding generations of respiratory bronchioles. Respiratory bronchioles are succeeded by alveolar ducts whose walls are formed by a musculoelastic mesh through which alveoli protrude outward. Alveolar ducts lead to the terminal, blind structures of the respiratory tree—alveolar sacs, which also are lined entirely by alveoli but in which there is no smooth muscle.

A variety of terms has been used for the same structures in the acinus, and the reader is referred to the excellent review by Pump.[8] It seems likely that divisions within the acinus follow a pattern of ir-

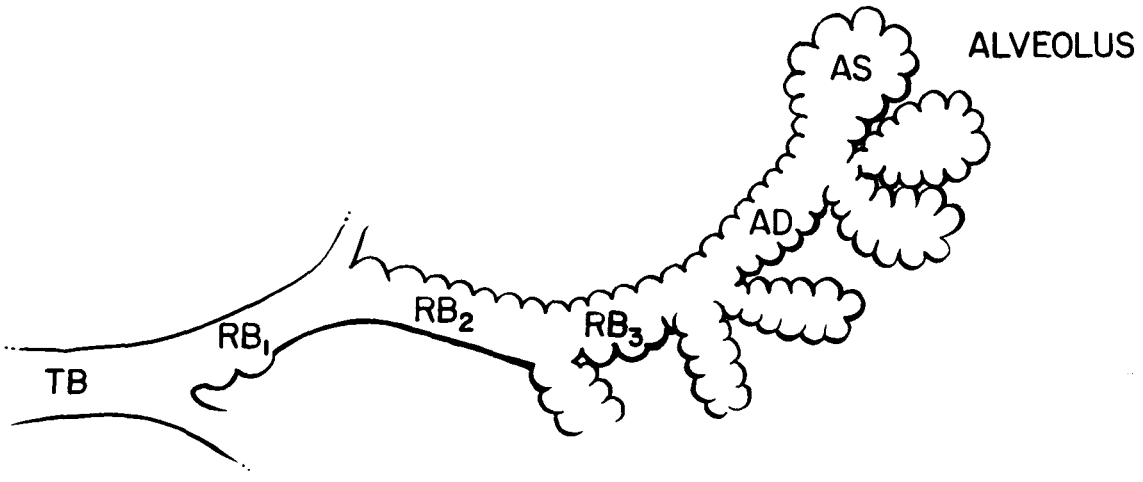

Figure 1–1. **Component Parts of the Acinus.** TB-terminal bronchiole. RB-respiratory bronchiole. AD-alveolar duct. AS-alveolar sac. (Thurlbeck, W. M.: Chronic obstructive lung disease. In: Sommers, S. C. (ed.), *Pathology Annual: Nineteen Sixty-eight.* Appleton-Century-Crofts, 1968, p. 377.)

regular dichotomy with axial and lateral pathways, as in the conducting system (*see* page 8). Thus, there may be from two to nine generations of respiratory bronchioles. The last respiratory bronchiole gives rise to a complex terminal spray of alveolar ducts and sacs in which dichotomous, trichotomous, or even quadrivial division occurs in irregular fashion. It can be estimated that there are about 400 alveolar ducts and sacs in an acinus and 18 last-order respiratory bronchioles arising from one terminal bronchiole. There are altogether about 14 million alveolar ducts and sacs, which contain the greater portion of the lungs' 300 million alveoli, each of which has a diameter of 250 to 300μ when fully inflated.[9] Alveoli are polygonal and have a continuous lining formed of flattened epithelial cells. In the sharp angles of the alveoli are plumper cells, with oval nuclei and granular cytoplasm. These corner cells, which are known also as type-II or septal cells, have many mitochondria, are metabolically highly active, and contain characteristic osmophilic inclusions which are thought to be the origin of the surface-active material that lines the alveolar surface.[10, 11] The total alveolar surface area is some 40 to 100 sq m, depending upon body size.[12] This area, in which gas transfer takes place, is extremely thin, and Schulz[97] has calculated that the thickness of the pathway from alveolar gas to the plasma layer of blood is 0.36 to 2.5μ—a small distance in terms of the diameter of a normal red blood cell.

The Lobule

The lobule is usually defined as the smallest discrete portion of the lung that is surrounded by connective-tissue septa, and corresponds to the "secondary lobule" described by Miller.[4] Since the septa are variable in size and extent, the size of the lobule is variable. It should not be confused with Miller's "primary lobule," which consists of an alveolar duct, its vessels, and the structures arising from them. The secondary lobule rarely is recognizable roentgenographically as a unit of structure; only when edema fluid or other pathologic tissue within the interlobular septa renders these structures visible as septal lines (B lines of Kerley; *see* Figure 4–52, page 242) can the volume of lung between two lines be recognized as a secondary lobule. It is obvious that such a limitation negates the usefulness of Miller's definition for purposes of pathologic-roentgenologic correlation.

To overcome this limitation, Reid and Simon[158] proposed an alternate definition, based on a subdivision of peripheral lung structure that is more constant and is recognizable both morphologically and roentgenographically. These authors pointed out that if one follows a bronchial pathway to the periphery of the lung in a well-filled bronchogram (Figure 1–2), a stage is reached at which branching of the parallel-walled pathway occurs every 0.5 to 1.0 cm. After three or four such branchings, an abrupt transition takes place in which the branching patterns occur at 2- to 3-mm intervals. These have been termed the "centimeter" and "millimeter" patterns, respectively. The centimeter pattern of branching represents small bronchi and bronchioles and the millimeter pattern relates to terminal bronchioles. Reid and Simon[158] proposed that the lobule be redefined as the cluster of three to five terminal bronchioles which form the millimeter pattern of branching at the end of a bronchial pathway (either axial or side branch), together with the respiratory tissue it supplies. As opposed to the conventional definition, theirs relates to a structure which is constant. We feel that this proposal possesses considerable merit: It designates a unit of lung tissue which is not only recognizable morphologically but also can be easily identified *in vivo* bronchographically.

Since each terminal bronchiole gives origin to a single acinus, and the Reid lobule comprises a cluster of three to five terminal bronchioles, the lobule is made up of three to five acini. Although variable in size, it measures approximately 1 cm in diameter in the adult.[158] If one divides a sphere 1 cm in diameter into three to five roughly equal parts, each part measures approximately 5 to 7 mm in diameter. Stated another way, division of a sphere 1 cm in diameter into units measuring 6 mm in diameter yields four to six units. These calculations agree with the original description by Loeshcke[7] in 1921, that the acinus measures approximately 6 mm in diameter, and with Reid's statement that the lobule

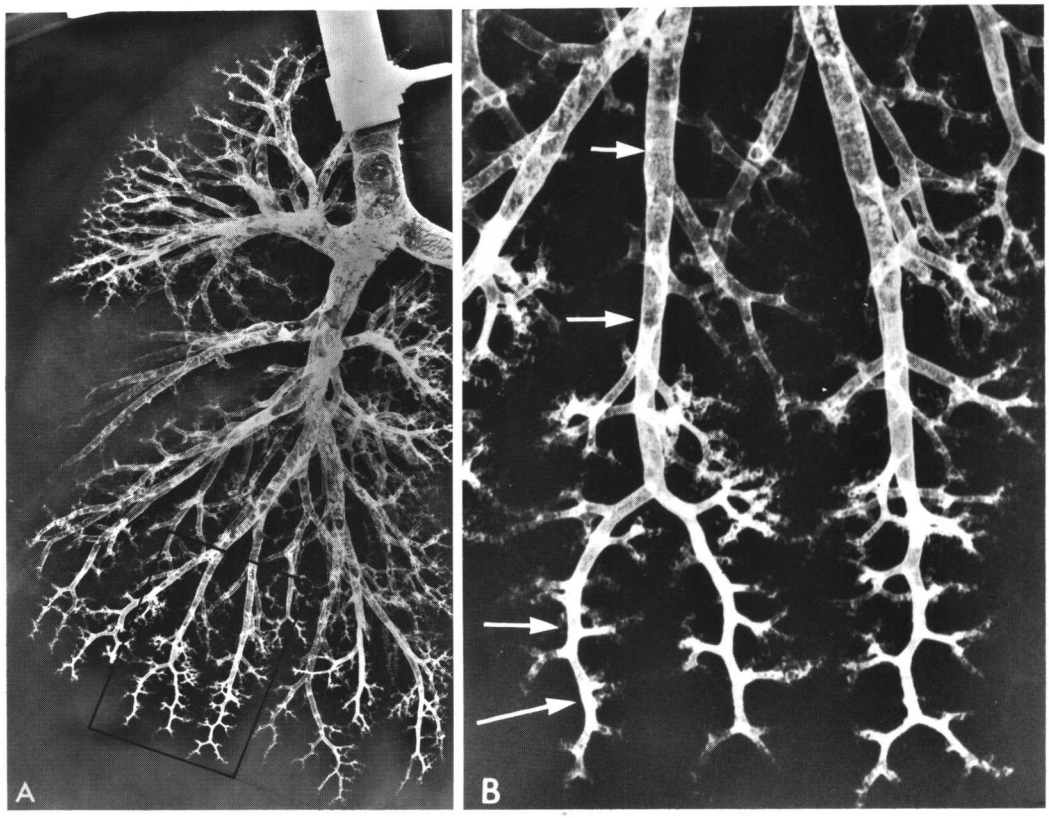

Figure 1–2. **The Pattern of Bronchial and Bronchiolar Branching.** *A*, Roentgenogram of an excised right lung following insufflation of particulate lead into the tracheobronchial tree. *B*, Magnified view of the peripheral airways of the lateral basal segment; upper arrows point to the "centimeter pattern" of branching, lower arrows to the "millimeter pattern." (Roentgenogram through the courtesy of Dr. James Hogg, Department of Pathology, McGill University.)

measures 1 cm in diameter and contains three to five acini, each approximately 6 mm in diameter.

Canals of Lambert

The distal portions of the bronchiolar tree, particularly the preterminal bronchioles, contain a number of epithelium-lined tubular communications with surrounding alveoli.[14] The physiologic significance of these structures is not known, but obviously they provide an accessory route for the passage of air directly from the bronchioles into alveoli.

Alveolar Pores

Although these are often called the pores of Kohn, von Hayek[15] attributes their original description to Henle. It is thought that they are present in the lungs of all mammals, though they may be relatively more common in some species. In the human lung, they are openings, or discontinuities, of the alveolar wall, about 10 to 15μ in diameter. The lung distal to an obstructed bronchus or bronchiole may be ventilated via an adjacent airway. This process, known as collateral air drift, is brought about, at least in part, by the pores of Kohn. The lung of the dog has no interlobular septa and, therefore, the lobar tissue is continuously interconnected.

Direct Airway Anastomoses

Martin[16] recently showed direct communications between "respiratory bronchioles" in the dog, measuring up to 120μ in diameter, which appeared to cross intersegmental planes. Although termed "respiratory bronchioles" by the author, the figures illustrate communications be-