SECOND EDITION

A DIAGNOSTIC APPROACH TO CHEST DISEASES

Differential Diagnoses Based on Roentgenographic Patterns

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TO
ELLEN AND ROBERTA,
WHOSE ENCOURAGEMENT MADE
THIS ENDEAVOR POSSIBLE,

AND TO OUR TEACHERS,
WHOSE CONCEPTS AND PRECEPTS
ARE EMBODIED THROUGHOUT THIS WORK.

Preface to the Second Edition

The reception accorded the first edition (1965) of this book demonstrated the need for a diagnostic approach of this kind, and the passage of the years has accentuated the advisability of a thorough revision more consistent with modern concepts. Much of the material has been completely rewritten to allow the updating of outmoded advice and to provide expanded discussions of certain topics. In addition, four new chapters have been added.

As stated in the first edition, this book is designed to provide a practical approach to the solution of diagnostic enigmas in pulmonary medicine. The standard textbook of medicine is basically a collection of monographs on individual disease entities and admittedly serves as an invaluable didactic tool and reference resource. In actual clinical practice, however, the patient presents himself to the physician, not with a diagnosis, but with a constellation of symptoms, physical signs, abnormal laboratory tests or radiological abnormalities which must be analyzed by the physician and synthesized with the results of new, appropriately-chosen diagnostic maneuvers in order to arrive at a label (diagnosis) which is sufficiently precise to permit rational therapy.

After the initial collection of data, the fundamental approach to diagnosis lies in the selection of the "problem"—a salient but encompassing clinical manifestation which serves as the framework for the construction of a differential diagnosis, and as a guideline for the selection of further diagnostic studies. Reflecting the dominant role that chest roentgenography has come to play in the practice of pulmonology,* the "problem" is frequently (but not invariably) a roentgenographic abnormality. An abnormal chest roentgenogram is often the sole evidence of the presence of disease but, even when symptoms and abnormal signs referable to the lungs are present, experience has shown that it is usually more productive to use the roentgenographic abnormality as the starting point in planning the diagnostic investigation. Most abnormal chest films can be categorized into roentgenographic patterns which lend themselves to this method of analysis.

Although the currently popular "problem-oriented" approach to medical practice is more concerned with the logical organization and recording of col-

* Pulmonology: the body of knowledge pertaining to pulmonary diseases; the practice of "chest" medicine and surgery (Waring).

lected data than with the patterns of diagnostic investigation, it is obviously compatible with, and analogous to, the method of differential diagnosis herein espoused. The concept of the use of the chest roentgenographic pattern as the initial focus for the diagnostic investigation was employed before the publication of the first edition of this book (Meschan, Simon) and since (Fraser and Paré, Reeder and Felson). As in the first edition, we have attempted to provide sufficiently detailed clinical correlations with the radiographic findings to give the reader a workable guide to the selection of the appropriate diagnostic strategy, and a summary of diagnostic criteria to indicate when the "end-point," the establishment of a working diagnosis, has been reached. Each chapter contains enough material that it can stand by itself with a minimum of cross referencing.

It is realized that occasionally a chest film cannot be easily categorized into any of the roentgenographic patterns described. To keep the discussions of differential diagnosis within manageable length and of practical value, we have often omitted from consideration the atypical and bizarre roentgen abnormalities occasionally manifested by certain common pulmonary diseases. It is believed, however, that the vast majority of "chest problems" are capable of resolution by the methods of analysis herein described.

This book was designed to provide a practical guide for the physician engaged in the clinical investigation of patients with undiagnosed bronchopulmonary diseases. It is not, in any sense, a manual of roentgenology, and most of the details of roentgenological interpretation have been deliberately omitted. In present day medical practice, the chest roentgenogram is usually interpreted by a specialist in diagnostic roentgenology and, in essence, this book tells the clinician how to proceed diagnostically after he has received the radiologist's report.

The diagnostic discussions pertain primarily to bronchopulmonary diseases in adolescents and adults, although some of the disease entities discussed occur more commonly in children. Pediatric chest problems are sufficiently unique that completely separate patterns of differential diagnosis would be required to deal adequately with the pulmonary diseases of infancy and childhood.

References are listed at the end of each chapter and have been rigorously pruned. Preference has been accorded to recent publications and to review articles which provide access to the classical contributions of present and past generations. Documentation throughout the text has not been attempted, although a specific author is occasionally cited where it seemed necessary to guide the reader to the appropriate reference. We cheerfully admit to the understandable (although not necessarily forgiveable) practice of including some references to papers in which we have paternal pride.

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Introduction

A. Diagnostic Management in Chest Disease

In medical practice, the term "management" is usually considered to pertain particularly to the selection and implementation of therapeutic programs. In an analogous fashion, the selection and implementation of the appropriate diagnostic studies to determine the nature of the disease process that is present can be categorized as "diagnostic management." Although our understanding of the theoretical basis for the optimal choice of diagnostic strategies is limited, certain fundamental axioms are generally accepted, and form the basis for the overall organization of this book and for the method of presentation within each chapter.

In general, the diagnostic process comprises four consecutive stages: (1) identification of the problem or problems which require investigation; (2) consideration of the diseases or syndromes which are potential causes of the specific problem, (3) the formulation and initiation of a diagnostic strategy in which diagnostic tests or maneuvers are employed in a systematic sequential fashion which will identify the specific etiology of the problem while minimizing delay, cost, pain, and risk; and (4) an assessment of the accumulated data to determine if a diagnosis has been established.

Identification of the Problem

In most instances of pulmonary disease, acquisition of a preliminary data base, comprising a history, physical examination, standard roentgenographic studies and "routine" hematological and urinary tests is advisable before a decision is made on the selection of the problem or problems which will form the reference point for further investigations. In the majority of cases, this will be a chest roentgenographic abnormality (i.e., a solitary pulmonary nodule, a pleural effusion, consolidation of one lobe of a lung, etc.). In many cases, the roentgenographic problem may be the only abnormality definable in an asymptomatic patient. Even when symptoms are present, it is our belief that selection of a significant roentgenographic abnormality as the focal point for diagnostic investigation is usually the most productive strategy. Symptoms and physical signs are not, of course, ignored, but are used to help determine the probabilities that certain diseases are the cause of the roentgenographic abnormality.

In some instances, a symptom or sign may be selected to form the basis for differential diagnosis. This is essential if a roentgenographic abnormality is absent, minimal, or clearly unrelated to the symptom. In emergency situations (acute respiratory failure, massive hemoptysis), or if the symptom or sign is a dominant clinical feature (moderate but not life-threatening hemoptysis), it is often advisable to regard the symptom as the "problem" for diagnosis, even if potentially significant roentgenographic changes are present. On occasion, it will be apparent that two seemingly unrelated but potentially significant pulmonary manifestations are present simultaneously, necessitating selection of *each* as a problem and resulting in the institution of two separate parallel diagnostic investigations.

Choosing Diagnostic Possibilities

Experience has shown that diagnostic failure due to selection of inappropriate diagnostic strategies is often due to a failure to consider all diagnostic possibilities. Once a problem has been identified and chosen, all possible causes of this clinical presentation should be enumerated (and preferably recorded).

On the basis of the information already available from the preliminary data base, it is often feasible to eliminate several of these diagnostic possibilities from further consideration if the probability that these conditions are the cause of the problem is extremely low. (However, additional clinical information may subsequently become available which will suggest that one or more of these "low probability" diseases should be reinstated in the list of possibilities.) This process of "pruning the diagnostic tree" will, of course, be continued as diagnostic testing provides further data.

After the initial listing and pruning of the diagnostic possibilities, it is often helpful to estimate the relative likelihood that each of the remaining diagnostic "candidates" is, in fact, the cause of the problem which is under consideration. This will assist in the selection of appropriate diagnostic tests.

Diagnostic Management

The formulation of diagnostic strategies will clearly be dominated by two considerations: (1) tests which are likely to prove or disprove the presence of "high probability" diseases will clearly have a high priority, and (2) if the "penalty" for failure to diagnose a disease is high (i.e., failure to diagnose pulmonary embolism may result in death), testing for such a disease will have a high priority even if the probability that it is the cause of the problem is low.

Desirable characteristics of a diagnostic test, roughly in descending order of importance, include low risk, pertinence to the problem under investigation, low discomfort, high specificity, high sensitivity, availability, minimal delay between performance and obtaining the results, and low cost. No single test has all these characteristics and the choice of one test over another will depend upon weighing these various factors.

In actual practice, several tests may be performed simultaneously. As the investigation progresses, the selection of further tests will be influenced by the changes in the diagnostic probabilities created by the data obtained from earlier tests. In some situations, therapeutic programs may be initiated concurrently with the diagnostic investigation. A "trial of therapy" is a useful diagnostic tool in certain circumstances. At times, it will be prudent to initiate therapy for a disease which carries a significant risk in the untreated state, even if a definitive diagnosis has not been established.

Establishment of the Diagnosis

The criteria by which one concludes that a specific diagnosis has been established will differ among different disease states. Some diseases (i.e., cancer) have a pathognomonic sign (i.e., positive biopsy) which, when detected, is diagnostic per se. In the case of certain other diseases, the diagnosis is considered to be established if the probability that a specific disease is the cause of the problem is sufficiently high, and the probabilities of all other diseases are sufficiently low. In certain conditions, the high probability is attained if certain criteria (major or minor) are present in combination. Finally, some diagnoses are reached by exclusion of other possibilities. For example, chronic bronchitis can be diagnosed as the cause of a chronic productive cough if other potential causes (such as lung tumor, tuberculosis and bronchiectasis) have been ruled out by appropriate diagnostic studies.

We realize that many accomplished clinicians will deny (or at least fail to recognize) that their diagnostic endeavors follow a pattern which resembles the one described. Certainly, the very act of analysis imposes a static pattern of formality which clearly has artificial qualities. In actual practice, the clinician, from the earliest stages of the collection of the initial data base, may be considering possibilities, weighing probabilities, and formu-

lating strategies; in fact he often uses these initial diagnostic impressions for guidance in obtaining a more comprehensive and pertinent history and to focus special attention on certain components of the physical examination. He may never formally choose a "problem" or prepare a complete "gamut" of diagnostic possibilities. We contend, however, that he often does this without consciously realizing it. Persistent failure to consider diagnostic possibilities other than the obvious one leads to "tunnel vision" and may have catastrophic consequences.

B. How to Use This Book

The content and organization of this book are designed to aid the physician in the diagnostic management of patients with undiagnosed bronchopulmonary diseases. After completion of the basic studies (history, physical exam, and initial laboratory tests), the diagnostician should review the chest roentgenogram, preferably in conjunction with a radiological colleague. He is then able to identify the problem (or problems) which require solution and around which he will construct his differential diagnosis. It is hoped that this book will provide him with the answers to the questions, "What diseases are capable of giving rise to this problem, which diagnostic procedures are appropriate in this situation, and how can I tell when the diagnosis is actually established?"

Part One deals with the diagnostic techniques of value in the investigation of patients with pulmonary diseases. It is designed for review and to minimize constant repetition throughout the remainder of the book. It is not an exhaustive account, and other works (listed in the references) should be consulted for details on the performance of the various procedures.

Part Two describes and illustrates the major roentgenographic patterns exhibited by abnormal chest films. When the physician is confronted with an abnormality in the chest x-ray, it is suggested that a review of Chapter 4 will enable him, in most instances, to assign his specific case to one of the roentgenographic categories described. He should then consult the appropriate chapter in Part Three for a description of the subsequent steps in the diagnostic investigation of the specific roentgenographic pattern in question.

In Part Three, a single chapter is devoted to each of the roentgenographic patterns described in Part Two. The material in each chapter is organized in the same fashion, beginning with a description of the disease processes that are likely to present the roentgenographic abnormality under consideration, and including certain clinical features of diagnostic value about each disease. The investigative procedures likely to prove helpful are then listed and briefly discussed. Each chapter concludes with a section termed "Diagnostic Management," in which the actual patterns of the investigation are indicated and in which the criteria for establishment of the final diagnosis are discussed.

In Part Four, a variety of problems are discussed which are not primarily radiological in character, although abnormalities are often present on the chest roentgenogram. These problems include symptoms (cough, dyspnea), a physical sign (hemoptysis), pathophysiological states (respiratory failure, comprised defenses against infections), and a syndrome (pulmonary infiltration with eosinophilia).

Contents

PART 1

AN OUTLINE OF DIAGNOSTIC TECHNIQUES IN BRONCHOPULMONARY DISEASE

CHAPTER 1

The History and the Physical Examination

Medical History in Differential Diagnosis of Chest Disease

General Considerations

It is a medical axiom that the clinical history, as it is provided by the patient himself and by other informants who have knowledge of the case, provides more assistance in diagnosis than any other single approach. It has been wisely said that if extensive diagnostic studies have failed to reveal the cause of the patient's illness, it is often more valuable to take the history again than to repeat the laboratory tests.

In some instances, the clinical history is so characteristic of a given disorder that the diagnosis can be made on the basis of the history alone, regardless of the outcome of the physical examination and the laboratory tests. In many instances, the clinical history will permit a tentative diagnosis to be made, subject to confirmation by roentgenological or laboratory tests. In most instances the history will at least provide some clues to assist in the selection of additional investigative studies.

In general, the clinical history is of somewhat less value in the study of chest diseases than in most other areas of internal medicine. It is a common occurrence for the chest roentgenogram to reveal evidence of extensive or serious lung disease in the absence of symptoms. Even when symptoms are present, they are often sufficiently nonspecific as to provide only a modicum of diagnostic assistance. Although the analysis of individual symptoms is often of little help, consideration of the symptom complex as a whole, particularly with regard to its sequential development, may be of considerable differential diagnostic value. Symptoms provide their greatest diagnostic assistance when considered in conjunction with the results of roentgenological and laboratory examinations.

The clinical history obtained from the patient may be recorded in the traditional fashion, beginning with the presenting complaint and continuing through the history of the present illness, previous illnesses, family history, and personal and social histories to a complete review of symptoms. This is followed by a complete description of the results of physical examination. The historical data can also be recorded in a *problem oriented* format. The delineation of the pertinent problems after acquisition of the initial data base comprises a value aid to differential diagnosis. Indeed, this book is organized so that each chapter (after Chapter 4) comprises the diagnostic management of a commonly encountered "problem" in chest disease. Such problems are often, but not

necessarily, a roentgenographic abnormality.

Certain aspects of the history require special consideration because of their frequency in patients with bronchopulmonary diseases, and specific inquiry must be made about these features if the information is not volunteered spontaneously. These cardinal symptoms of bronchopulmonary disease are therefore discussed in some detail.

Cardinal Symptoms of Bronchopulmonary Disease

Cough. Cough occurs in such a wide variety of respiratory diseases that it has relatively little differential diagnostic value. Indeed, in neurotic subjects it may be a very prominent symptom in the absence of respiratory tract disease. Conversely, the patient may minimize or even deny the presence of cough despite the fact that he coughs throughout the entire interview.

Cough is an explosive expiratory maneuver which can be performed voluntarily or may occur as a reflex reaction to irritation. The purpose of the cough is to clear the airways of foreign material or excess mucus and it results from stimulation or irritation of the upper or lower airways from the larynx to the terminal bronchioles. Alveolar disease does not usually cause cough unless there is an associated bronchial irritation.

An effective cough requires efficient glottic closure to develop the "tussive squeeze," and adequate chest muscle strength and open airways to permit the "bechic blast." The cough reflex may be markedly diminished by narcotic or sedative drugs, and the effectiveness of the cough is reduced by chest pain, muscular weakness or paralysis, and laryngeal disorders preventing adequate glottic closure (i.e., tracheostomy). Excessively thick secretions and diseases causing diffuse airway obstruction will markedly reduce the attainable expiratory flow rates, and the cough will be weak and often nonproductive.

In most patients, cough is due to such relatively innocuous or mundane conditions as chronic postnasal drip, acute upper respiratory tract infections, chronic bronchitis, and the inhalation of cigarette smoke. The significance of the symptom of cough should *never* be minimized, however; an adequate explanation should be sought in all cases. A chest x-ray should always be obtained, but a negative x-ray does not rule out the possibility of serious disease. A persistent unexplained cough is an indication for bronchoscopic examination.

Certain characteristics of the cough may be of diagnostic value. A loud, harsh, hacking cough usually originates from irritation in the larynx, trachea, or main bronchi. A painful cough indicates pleural involvement or irritation, a dorsal spine lesion with root pain, or chest wall disease (including the "tussic rib fracture" secondary to excessive coughing). Coughing that occurs during or immediately after swallowing may indicate the presence of a tracheoesophageal fistula or a disorder of swallowing that leads to aspiration. An emetic cough, which results in expectoration of previously ingested food stuffs, suggests hiatal hernia, pharyngeal diverticula, or recurrent aspiration. A severe prolonged paroxysm of coughing may cause retching, or syncope. Cough is commonly present in asthmatic patients and is ordinarily associated with wheezing respirations. The diagnostic management of cough is considered in greater detail in Chapter 28.

Expectoration. The term expectoration implies that the material produced by coughing has originated within the chest. Postnasal drainages cleared from the throat in the morning after awakening may be difficult to differentiate from true expectoration. Children and women often swallow any sputum that is brought up into the pharynx. The physical characteristics of the sputum are of some diagnostic assistance. Viral bronchitis usually causes a thin, mucoid sputum, whereas bacterial infections of the lung or bronchi are associated with thick, discolored "purulent" sputum. In patients with chronic bronchitis and emphysema, the change from mucoid to purulent sputum may be the only clini-

cal evidence that an acute infection or flareup has developed. In lung abscess or bronchiectasis, several ounces of purulent, foul smelling sputum may be produced daily. Bronchorrhea (the daily expectoration of large quantities of watery sputum) is very suggestive of the presence of an alveolar cell carcinoma of the "diffuse" variety. A history of expectoration of multiple, small or large, hard, gritty, calcific masses is virtually pathognomonic of broncholithiasis. In acute pulmonary edema, the sputum is characteristically frothy, pink, and profuse, often welling up into the throat with minimal coughing. Patients with asthma usually cough during the paroxysmal attacks and at times the asthmatic may expectorate viscid sputum which exhibits a branching pattern and represents actual casts of small bronchi and bronchioles. Patients with allergic bronchopulmonary aspergillosis may cough up large, firm, brown mucoid plugs which are solid and rubbery. In pneumococcal pneumonia the sputum is viscid and brown and in Friedländer's pneumonia it is often sticky and slimy. In patients with pseudomonal infections, the sputum is often green in color.

Hemoptysis. Hemoptysis is a relatively common symptom which is of great importance for two reasons: it is often a manifestation of serious pulmonary disease, and it usually arouses great apprehension in the patient and causes him to seek medical consultation. A wide variety of bronchopulmonary diseases may give rise to hemoptysis. Common causes include chronic bronchitis, bronchogenic carcinoma, bronchiectasis, mitral stenosis, chest trauma, and pulmonary infarction. The differential diagnosis of hemoptysis is discussed in detail in Chapter 27.

Dyspnea. Shortness of breath is a frightening symptom which usually induces the patient to seek medical advice promptly. The pathogenesis of the abnormal sensation is poorly understood, and the correlation of the severity (or even the presence) of the symptom with the clinical, radiological, and physiological findings is far from exact.

A sensation of breathlessness is a common complaint in neurotic subjects who have no pulmonary disease. Such people often describe the symptom as a feeling that "the air isn't able to get all the way down and doesn't seem to do me much good." The patient attempts, consciously or subconsciously, to compensate by breathing more deeply, and the characteristic features of the hyperventilation syndrome may then supervene. Anxiety induced hyperventilation may also occur in patients with organic dyspnea from cardiac or pulmonary disease and it presents an extremely difficult diagnostic and therapeutic problem.

Certain features of dyspnea may provide diagnostic assistance. The characteristic expiratory wheeze is virtually pathognomonic of diffuse obstructive disease of the smaller airways (asthma, asthmatic bronchitis, emphysema), but may occur in some cases of cardiogenic pulmonary congestion (cardiac asthma) and can be simulated voluntarily by some people with normal lungs and airways. Wheezing respiration may be due to a localized obstruction of a major bronchus (tumor, stricture, foreign body) but the unilaterality of the wheeze is usually evident at physical examination. An inspiratory stridor indicates partial obstruction at the laryngeal or tracheal level. Orthopnea (dyspnea increased by lying flat) is very characteristic of cardiogenic pulmonary congestion but is sometimes a feature in asthmatic and emphysematous patients. Paroxysmal wheezing unrelated to physical exertion or posture is a typical manifestation in bronchial (allergic) asthma.

An *acute* onset of dyspnea in a previously well person suggests such possible causes as pulmonary embolism, acute left ventricular failure, pneumonia, spontaneous pneumothorax, foreign body aspiration, acute laryngeal edema, croup, and massive pulmonary hemorrhage. *Recurrent* episodes of acute dyspnea suggest asthma or the paroxysmal nocturnal dyspnea of chronic left ventricular failure. Aggravation of dyspnea in a patient with emphysema suggests a superimposed acute bronchial infection.

Chronic progressive dyspnea is a char-