RADIOLOGY OF THE PEDIATRIC CHEST

CLINICAL AND PATHOLOGICAL CORRELATIONS

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Notice

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To my wife Lynne
my boys David, Robert, James—
this book is affectionately dedicated

Preface

The writer's only responsibility is to his art. He will be completely ruthless if he is a good one. He has a dream. It anquishes him so he must get rid of it. He has no peace until then. Everything goes by the board: honor, pride, decency, security, happiness, all, to get the book written. If a writer has to rob his mother, he will not hesitate; the "Ode on a Grecian Urn" is worth any number of old ladies.

William Faulkner, 1956

Of two things I am certain: (1) this book will never be confused with "Ode on a Grecian Urn," and (2) it could never have been written without the help of several, very reliable "ladies." Nevertheless, Faulkner's words have fueled my furnace for the last year, and I quote him with reverence, respect, and considerable tongue in check.

Physicians who care for children with respiratory tract illness must recognize the roentgenographic expression of a host of disorders. Radiologists, who wish to fulfill their role as consultants, must be familiar with the clinical manifestations of these diseases as well as their radiographic presentations. Rapid advances in our knowledge of pathophysiology and therapy, as well as the explosion of radiographic imaging technology, compound the task of each.

My major purpose is to bring together the clinical and radiologic features of pediatric chest diseases in a form that is comprehensible to the most uninitiated medical student, yet valuable to the sophisticated pediatric radiologist. The material is presented in a descriptive manner; struc-

tured algorithms and "cookbook" recipes are avoided since clinical presentations, available facilities, local customs, and individual expertise are rarely comparable. Well-informed clinicians, radiologists, and pathologists, trained to assess cases on their individual merits, should find the most logical path to the proper diagnosis and treatment regimen.

Section IV, "Roentgenographic Patterns in Pulmonary Disease," is devoted to four patterns that I have found reliable for pediatric chest interpretation. The "pattern" recognition and gamut list methodologies, so dear to medical students and residents, often suffer the danger of becoming the proverbial square pegs in round holes. This method should not be substituted for careful observation and description, logical analytical discussion, and reasonable differential diagnosis.

Discussions and illustrations of the associated pathologic processes are placed wherever needed to expand and clarify the behavior and nature of these abnormalities. My fascination with pathology reaches back to my monumental medical school chairman of pathology, Dr. Edward A. Gall, who continues to prod me from his grave in Cincinnati, Ohio. Time and distance offer no escape from him.

Without doubt, the most significant and rare stroke of genius was my decision to enlist the talents of Dr. Mervyn D. Cohen, of the James Whitcomb Riley Children's Hospital. With hardly a twitch of his whiskers, he accepted the challenge and, within several months, delivered three beautifully composed and illustrated chapters on chest imaging in children. His broad experience with computed tomography and his pioneering role in magnetic resonance imaging are unexcelled, and I value his contribution greatly.

As I close this prologue, my thoughts wander back to my roots in the Cincinnati College of Medicine, the Cincinnati Children's Hospital, and Dr. A. Ashley Weech, that marvelous pediatrician, educator, and humanitarian, whose legacy of inspiration lives within all of us who called him "Pappy." His words, his wisdom, his way with children, are entwined within these pages.

ALVIN H. FELMAN

Acknowledgments

Any attempt to acknowledge the many individuals who contributed to the production of this book is doomed to failure at the outset. Nevertheless, I cannot let the occasion pass without at least some effort.

The support of the photographic department of the University Hospital, headed by Stephen Englert, was willing and prompt, in spite of my many trying demands. Linda Laughton, a secretary of unexcelled skill, typed the original draft with a promptness and accuracy rarely found.

Many of the chapters were reviewed by Davey Volkhardt, editorial consultant for the University of Florida, Jacksonville campus. Davey is a writer and poet in her own right; a true woman of letters, the likes of whom I have not encountered since my days in high school English class. If any participles have been left dangling, or if my assault on the English language has at times exceeded the bounds of credulity, it is the result of my own stubborn ignorance and should cast no shadow on Davey's knowledge or the futile tenacity with which she so often tried to restrain me.

My wife, Lynne, was a constant source of support and inspiration; this book could never have been completed without her. She sat by my side in front of our word processor during the endless, tedious editing process and tried to keep me from writing foolishness. I would like to believe she was successful, but, in truth, her task was insurmountable.

Dr. Ronald Rhatigan, medical director and chairman of the Department of Pathology, was a willing and generous patron of this project, as

were his associate pathologists, Drs. Jeffrey Goldstein and Carmela Monteira. Only a pathologist can experience the sheer terror and disruption of a radiologist in the pathology department. They suffered my bullish intrusions with patience and forbearance, even in the midst of their "frozen" sections.

Dr. William Donnelly of the University of Florida provided much of the pathologic material for the previous edition that has been carried over into this version. I again owe him my gratitude for his past contribution to this effort.

My thanks and appreciation are also expressed to the Department of Radiology chairman, Dr. Frederick Vines, and my colleagues and house officers, for their support and encouragement.

The unselfish willingness of so many of my colleagues in radiology to share their experiences is evident from the unique and beautifully documented cases that they contributed. Other authors whom I contacted were equally generous. I would like to recognize their contributions by listing their names below.

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Contents

Section I

Congenital Abnormalities of the Larynx, Trachea, Esophagus, Bronchi, and Lungs

Developmental anomalies of the larynx, trachea, esophagus, bronchi, and lungs usually produce severe neonatal distress in the first critical hours or days of life. If undiscovered and untreated, irreparable damage may result, most often from aspirated oral feedings or gastric contents. Approximately one-half of children born with these deformities have associated congenital anomalies, and half of these are fatal.

The three chapters in this section will focus on the radiographic expression of some of these anomalies. Effort is made to correlate the clinical and radiographic pictures with the embryologic etiologies. Whenever possible, pathologic specimens are included for emphasis, clarity, and interest.

Chapter 1 summarizes the major congenital anomalies caused by abnormal or incomplete septation of the primitive foregut. In general, these manifest as atresias, fistulas, and/or defects in the tracheoesophageal septum, or "party wall."

Chapter 2 considers congenital lesions that result from abnormal or aberrant lung bud development. Among these are ectopic bronchial connections, underdeveloped lungs, "sequestered" lobes, and cystic formations.

Chapter 3 contains a heterogeneous group of lesions in which the major aberrations involve parenchymal lung structures, such as alveoli,

blood vessels, supporting stroma, and peripheral bronchial tree. It is difficult to trace the teratogenic roots of these lesions; some may actually be acquired rather than congenital.

Chapter 1

Abnormal Tracheoesophageal Development

1. Abnormal *differentiation* of the primitive foregut into the trachea, larynx, and/or esophagus
   a. Tracheal agenesis-atresia
   b. Esophageal atresia with or without tracheoesophageal fistula
   c. Laryngotracheoesophageal cleft, esophagotrachea, H(N) fistula
2. Abnormal *maturation* and development of the trachea and larynx after normal differentiation has occurred
   a. Tracheomalacia
   b. Laryngomalacia
   c. Tracheal stenosis
   d. Tracheobronchomegaly

Abnormal Differentiation of the Primitive Foregut

Tracheal Agenesis-Atresia

Tracheal atresia is a rare anomaly that results when the trachea fails to develop; air reaches the lungs through esophageal communications. Floyd et al.[1] have classified tracheal agenesis-atresia into three types; type II is the most common (Figure 1-1). Pulmonary abnormalities in association with tracheal agenesis are unusual, but associated cardiac, gastrointestinal, and genitourinary anomalies are frequent.[2]

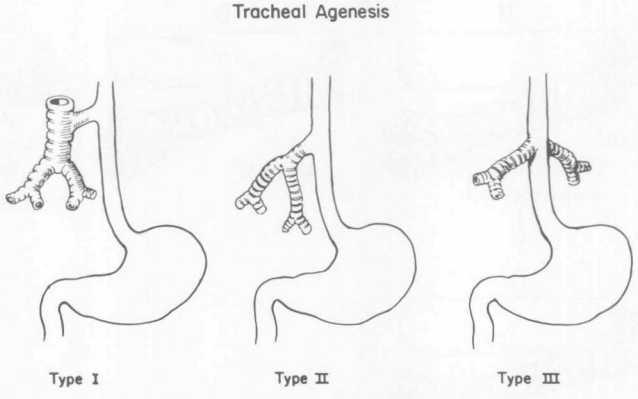


Figure 1-1 Tracheal agenesis. Type II is most common. (Adapted from Floyd et al.[1])

Because of the associated congenital defects, tracheal atresia may be considered within the group of anomalies referred to as the *VATER association* (*v*ertebral, *a*nal, *t*racheal, *e*sophageal, and *r*enal defects).[3] *VACTERL association* is a term occasionally used to signify the additional presence of cardiac and limb abnormalities.

While almost universally fatal, sporadic cases of tracheal atresia have been reported in which surgical repair has prolonged survival from days to several months and beyond.[4,5]

Embryology

Tracheal agenesis results from abnormal embryogenesis between the twenty-first and twenty-fourth day of gestation. During this period, the primitive foregut differentiates into the trachea and esophagus (Figure 1-2). Simultaneously, the future airway arises as a midline, ventral diverticulum, or lung bud. In the ensuing 10 days, the lung bud divides, elongates, and branches while the lateral esophageal grooves and tracheoesophageal septum complete the separation of the primitive foregut into trachea and esophagus. Normally, these structures are completely developed by 34 days, and laryngeal differentiation follows shortly thereafter.

Bremer[6] has suggested that aberrant deviation of the tracheoesophageal septum causes esophageal and/or tracheal atresia, with or without fistulas. Dorsal deviation of the septum leads to esophageal atresia (Figure 1-2D), with or without tracheoesophageal

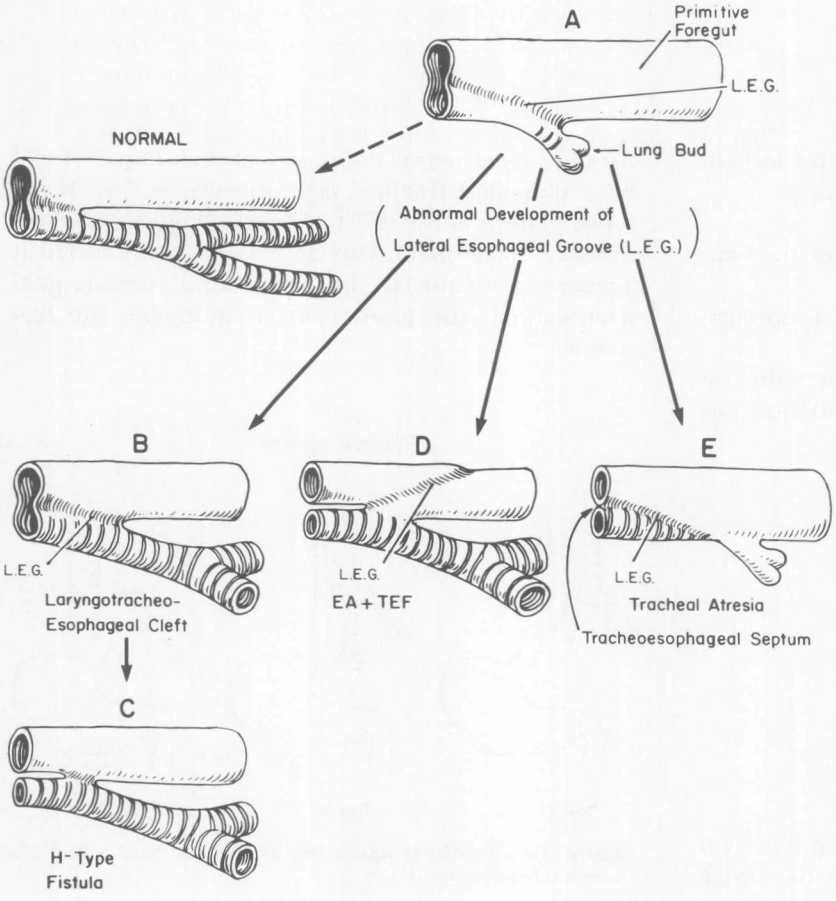


Figure 1-2 Diagram of normal and abnormal tracheoesophageal differentiation. A. At 21 to 24 days of gestation, the lateral esophageal grooves (LEG) appear and begin the separation of the foregut into trachea and esophagus. This process proceeds in a caudocranial direction until the entire tracheoesophageal septum is developed. As the foregut elongates, the ventral lung buds arise simultaneously, begin to branch, and ultimately give rise to the tracheobronchial tree. **B.** Failure of normal development and migration of the lateral esophageal grooves result in variable degrees of communication between the trachea and esophagus. (See Fig. 1-9.) **C.** The H(N) fistula probably results from a localized developmental defect in the tracheoesophageal septum (see Figure 1-3E). **D.** Localized overgrowth of the lateral esophageal grooves or dorsal deviation of the tracheoesophageal septum leads to the common form of esophageal atresia and tracheoesophageal fistula (see Figure 1-3A). **E.** Ventral deviation of the tracheoesophageal septum or abnormal development of the lateral esophageal groove may result in tracheal atresia (see Figure 1-3D). The simultaneous development of a caudal lung bud may give rise to esophagotracheal fistula or esophageal lung (see Figure 2-1). Oblique deviation of the tracheoesophageal septum is probably responsible for other combinations of abnormal bronchoesophageal attachments (see Figure 1-3B, C, F).

fistula, whereas ventral deviation produces tracheal atresia (Figure 1-2E). During this complicated developmental process, supernumerary or aberrant lung buds may give rise to esophageal bronchi or other anomalies (sequestration? bronchogenic cyst?). These anomalies of abnormal lung bud development are considered in more detail in Chapter 2. Disturbances in the sequence of septal deviation, esophageal elongation, and lung bud development probably account for the clinical variations of tracheal and esophageal atresias depicted in Figure 1-3. In addition to Bremer's theory of embryogenesis, other authorities have suggested that hypertrophic development of the lateral esophageal grooves plays a part in the genesis of these complex anomalies.[7]

Clinical Symptoms

Infants with tracheal agenesis present with cyanosis, severe respiratory distress, and lack of audible cry. Inability to intubate the trachea is a consistent feature. Hydramnios is frequently present, but oligohydramnios occurs with accompanying renal dysgenesis. Associated external congenital abnormalities may be evident, and, if the infant survives, additional developmental defects may become manifest. Tracheal intubation is impossible; the endotracheal tube usually enters the esophagus. Nevertheless, some oxygen delivered through a tube in the esophagus will reach the lungs via tracheoesophageal communication. Thus, life is sustained for a few hours or days. The larynx is usually well developed.

Radiologic Findings

In infants who have tracheal atresia and survive long enough to have film studies, the lungs may appear surprisingly well aerated.[2] Within a short time, generalized opacity, collapse, and pneumothoraxes appear (Figure 1-4A). The tracheal air column is not visible (Figure 1-5A), and endotracheal tubes usually are displaced into the esophagus and stomach. Contrast esophogram usually confirms the presence of bronchoesophageal communication (Figures 1-4B and 1-5C). The cardiac silhouette may or may not be abnormal, depending upon the presence and type of congenital heart defects, but the heart may appear normal at birth even in the presence of complex cardiac disease. Vertebral deformities occasionally occur in association with tracheal atresia.

Esophageal Atresia and Tracheoesophageal Fistula

The combination of esophageal atresia and tracheoesophageal fistula (EA/TEF) occurs once in every

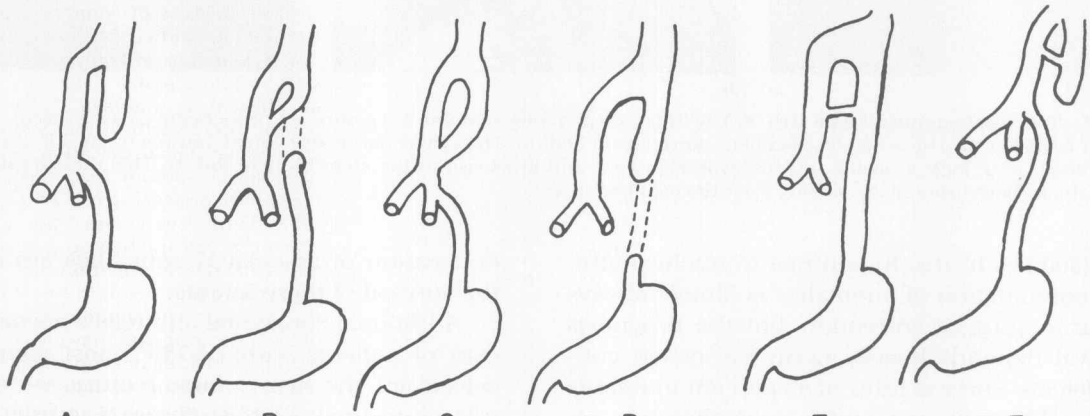
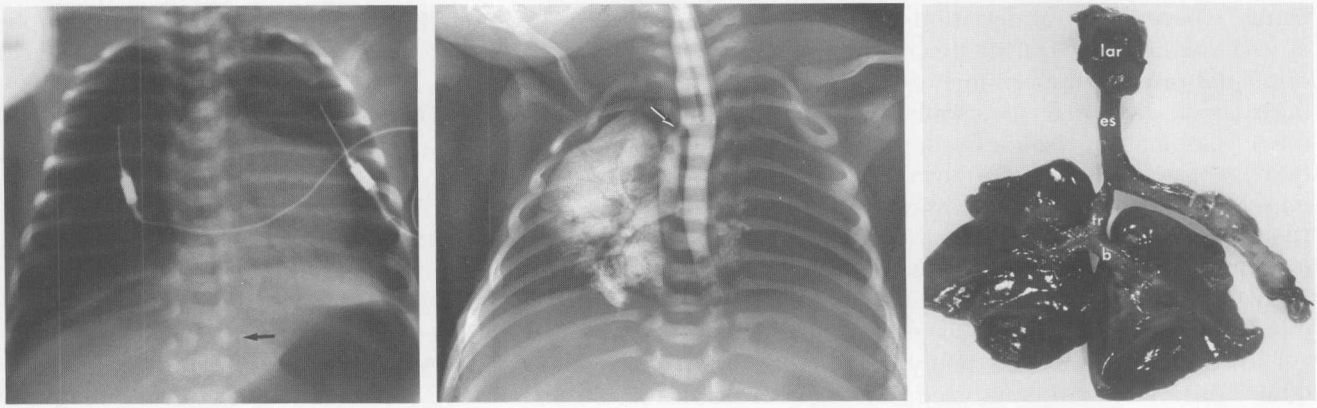
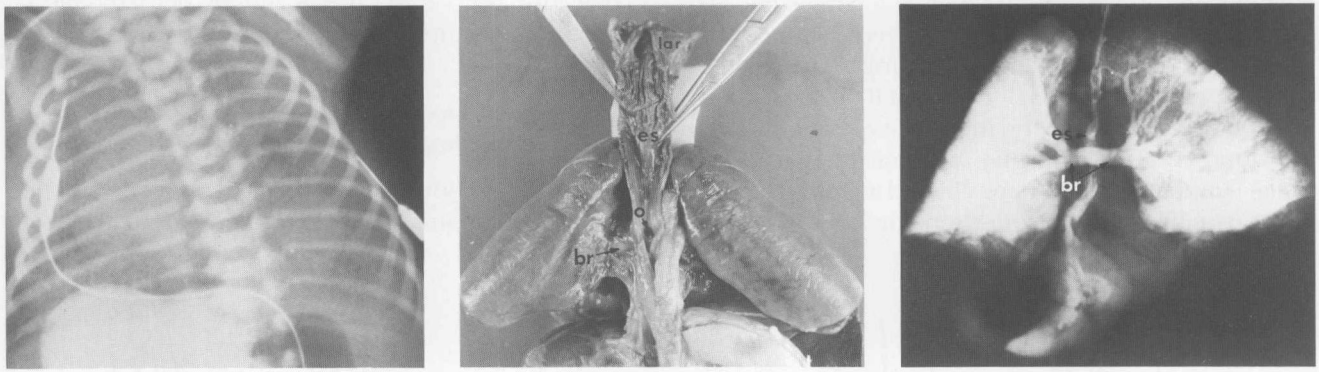


Figure 1-3 Diagram of tracheoesophageal abnormalities. Type A shows the most common abnormality, accounting for approximately 95 percent of cases.



A B C

Figure 1-4 Tracheal agenesis (type I). This 2200-g infant could not be intubated. Tracheostomy was performed, but the patient died at approximately 6 h of age. Imperforate anus and polyhydramnios were additional complications. **A.** The only film study obtained shows bilateral pneumothoraxes and pneumomediastinum. There is gas in the stomach. *Note:* vertebral malformations (arrow). **B.** A postmortem contrast injection shows the tracheoesophageal connection (arrow). **C.** Autopsy specimen: The trachea (tr) originates directly from the midesophagus (es). The tracheal segment is narrow at its origin, but the tracheal rings in this region and distally are unremarkable. The larynx (lar) is well formed. (b) = bronchus.



A B C

Figure 1-5 Tracheal agenesis (type III). **A.** The lungs are generally opaque, but central air bronchograms are present. The tracheal air column is absent. **B.** The autopsy specimen, viewed from behind. The larynx (lar) is developed, but the trachea is absent. A single orifice (o) arises anteriorly from the opened esophagus (es) and gives rise to the stem bronchi (br). **C.** This radiograph of injected specimen shows both bronchi (br) arising from the esophagus (es).

3000 to 3500 live births. In contrast to tracheal atresia, this combination of anomalies is almost always amenable to surgical correction, but the prognosis for survival depends heavily upon associated congenital defects. Survival rates of 88 percent in infants with no additional congenital anomalies are reported.[8] Early recognition of this abnormality, accurate evaluation of the anatomic defect, and thorough delineation of associated anomalies are critical for the survival of these infants.

Additional congenital anomalies occur in 50 percent of patients with EA/TEF; most commonly involved are the heart, anus, rectum, vertebrae, and gastrointestinal and genitourinary tracts (VATER-VACTERL associations). Thymic and parathyroid abnormalities also accompany EA/TEF, as in Di-