

Other diagnostic procedures may include chest films showing infiltrates that may involve a single lobe (lobar pneumonia from staphylococci) or may be more diffuse as in the case of bronchopneumonia (usually streptococci). Physical examination, including percussion and auscultation of the chest, may reveal signs of lung consolidation such as dullness, inspiratory crackles, or bronchial breath sounds.

TREATMENT. The primary treatment for bacterial and mycoplasmic forms of pneumonia is antibiotic therapy along with rest and fluids. Treatment with specific antibiotics is based on the history; whether the pneumonia was community-acquired, hospital-acquired, or extended care facility-acquired; and on the medical status and overall condition of the client (e.g., otherwise healthy or debilitated). Airway clearance techniques (formerly, chest physical therapy, pulmonary physical therapy, and pulmonary hygiene) may aid in clearing purulent sputum.

Fungal pneumonia is treated with antifungal drugs such as itraconazole or amphotericin B. Viral pneumonia is treated symptomatically unless secondary bacterial pneumonia develops. Hospitalization may be required for the immunocompromised client.

A vaccine is recommended for everyone age 65 years or older; for people with chronic disorders of the lungs, heart, liver, or kidneys; for individuals with poorly controlled diabetes mellitus; and for those with a compromised immune system or confined to a long-term care facility. Immunization can provide protection from pneumococcal disease for a period of 3 to 5 years in over 80% of vaccinated persons. A pneumococcal conjugate vaccine for routine use in infants and in high-risk children effective against invasive pneumococcal disease and to a lesser degree against otitis media and pneumonia has been licensed for use in the United States.^{314,386}

The pneumonia vaccine has been successful in reducing penicillin-resistant *S. pneumoniae* by 81% in infants and 49% in the elderly between 1999 and 2004. Because pneumonia is a common complication of the flu, the U.S. Centers for Disease Control and Prevention (CDC) recommends annual flu vaccinations as well.

PROGNOSIS. Community-acquired pneumonia remains a common and serious clinical problem despite the availability of potent antibiotics and aggressive supportive measures. Hospital-acquired pneumonia (HAP) has an even higher mortality rate. Pneumonia ranks seventh among the causes of death in the United States and currently accounts for almost 40% of hospital deaths; 90% of those fatalities occur in people over age 65 years, largely a result of coexisting medical problems that weaken the immune system.

Highly effective prevention and treatment methods can improve survival and reduce the likelihood of developing pneumonia, but one-half of older adults do not get vaccinations that could cut the death rate in one-half. The *Healthy People 2010* Objective 1-9c is to reduce hospitalization for immunization-preventable pneumonia to 8 per 10,000 in persons aged 65 years or older.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-3

Pneumonia

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

The CDC recommends adherence to standard precautions for clients with pneumonia. At the very least, careful handwashing by all personnel involved is essential for reducing the transmission of infectious agents. Adequate hydration and pulmonary hygiene, including deep breathing, coughing, and airway clearance techniques, should be instituted in all clients hospitalized with pneumonia. Ventilatory support and supplemental oxygen may be needed to maintain adequate gas exchange in severely compromised clients.

Preventive measures are important and include early ambulation in postoperative clients and postpartal women unless contraindicated. Proper positioning to prevent aspiration during the postoperative period and for all people who are immobilized or who have a poor gag reflex is important.

Occasionally, a lower lobe infection can irritate the diaphragmatic surface so that pain referred to the shoulder is the presenting symptom. For the client with a known diagnosis of pneumonia, the breathing pattern and the position assumed in bed can indicate the client's discomfort, reveal tachypnea, and demonstrate splinting of the chest to minimize pleuritic pain (i.e., lying on the affected side reduces the pleural rubbing that often causes discomfort).

Lobes affected by pneumonia will remain vulnerable to further infection for some time, especially in the bedridden, debilitated, or neuromuscularly compromised population. Airway clearance techniques are not usually helpful in adults with uncomplicated pneumonias. However, the client, family, or caretakers should be instructed in breathing exercises and a positional rotation program with frequent positional changes to prevent secretions from accumulating in dependent positions and to optimize ventilation/perfusion matching.

Pneumocystis Carinii Pneumonia

Definition, Etiology, and Risk Factors

Pneumocystis carinii pneumonia (PCP) is a progressive often fatal pneumonia. The origin of the organism is unknown. It is possibly acquired from the environment; infected humans; or animals, fungi, or protozoa. Other people at risk for the development of PCP include anyone who is immunosuppressed for organ transplantation, by

chemotherapy for lymphoma or leukemia, by steroid therapy, or by malnutrition.

Previously, the majority of people with AIDS developed PCP during the course of their illness, but this is much less common now with pharmacologic prophylaxis. PCP has been shown to be the first indicator of conversion from human immunodeficiency virus (HIV) infection to the designation of AIDS. In a retrospective 10-year analysis of people with PCP, 6 out of 18 patients were HIV positive.²⁰⁸

Pathogenesis and Clinical Manifestations

Infection begins with the attachment of the *Pneumocystis* trophozoite (the feeding stage of a sporozoan parasite) to the alveolar lining cell. The trophozoite feeds on the host cell, enlarges, and transforms into the cyst form that ruptures to release new trophozoites, repeating the cycle. If the process is uninterrupted by the immune system or antibiotic therapy, the affected alveoli progressively fill with organisms and proteinaceous fluid until consolidation disrupts gas exchange, slowly causing hypoxia and death.

The physiologic response to PCP includes fever, impaired gas exchange, and altered respiratory function. Symptoms of PCP develop slowly and present as fever and progressive dyspnea, accompanied by a nonproductive cough. Fatigue, tachypnea, weight loss, and other manifestations of underlying immunosuppressive disease may be present and worsened as a result of the increased metabolic demands.

MEDICAL MANAGEMENT

DIAGNOSIS, TREATMENT, AND PROGNOSIS. Molecular techniques in the laboratory play an essential role in the microbiologic diagnosis of pneumonia in the immunocompromised person. Other diagnostic tools may include fiberoptic bronchoscopy to obtain respiratory specimens for testing and chest radiograph. Diagnosis is important because effective pharmacologic treatment is available. Thanks to a worldwide collaborative effort among health care providers, academia, governments, and industry, our knowledge about and treatment of infection caused by HIV have evolved from palliative care to the use of a chronic disease model where survival is measured by decades, not months or years.¹⁹⁷

Although pulmonary disease remains a major problem for people with HIV, prophylaxis against opportunistic infection in people with HIV has cut morbidity and mortality rates by 80%.²¹ The increasing seroprevalence of HIV among women of reproductive age, the risks of vertical transmission of HIV, and the fact that PCP is the most common infection seen in people progressing to AIDS have led to recommendations for routine prenatal HIV infection counseling and testing.

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Carefully adhere to standard precautions to prevent contagion. Teach the client energy conservation techniques (see Box 9-8), as well as deep-breathing exercises. Expiratory air-flow reductions that persist after the acute infection resolves have been documented in cases of PCP (and bacterial pneumonia) in HIV-infected individuals. The clinical implications of these changes are unknown but may contribute to prolonged respiratory complaints and compromise in HIV-infected individuals who have had PCP.²¹⁰ See also Special Implications for the Therapist: Pneumonia in this chapter.

Pulmonary Tuberculosis

Definition

Tuberculosis (TB), formerly known as consumption, is an infectious, inflammatory systemic disease that affects the lungs and may disseminate to involve lymph nodes and other organs. TB is caused by infection with *Mycobacterium tuberculosis* and is characterized by granulomas, caseous (resembling cheese) necrosis, and subsequent cavity formation. Latent infection is defined as harboring *M. tuberculosis* without evidence of active infection; active infection is based on the presence of clinical and laboratory findings.

Overview

TB may be primary or secondary. The first or primary infection with the tubercle bacillus is usually asymptomatic and almost always (99%) remains quiet after the development of a hypersensitivity to the microorganism. The primary infection usually involves the middle or lower lung area with lesions consisting of exudation in the lung parenchyma. These lesions quickly become caseous and spread to the bronchopulmonary lymph nodes, where they gain access to the bloodstream and predispose the person to the subsequent development of chronic pulmonary and extrapulmonary TB at a later time.

Secondary TB develops as a result of either endogenous or exogenous reinfection by the tubercle bacillus. This is the most common form of clinical TB. Reactivated TB usually causes abnormalities in the upper lobes of one or both lungs. In the United States, development of secondary TB is almost always the result of endogenous reinfection that occurs when the primary lesion becomes active as a result of debilitation or lowered resistance.

Incidence

Despite improved methods of detection and treatment, TB remains a global health problem with the highest rates in Southeast Asia, sub-Saharan Africa, and eastern Europe.

SPECIAL IMPLICATIONS FOR THE THERAPIST

15-4

Pneumocystis Carinii Pneumonia

PREFERRED PRACTICE PATTERNS

6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders (AIDS/HIV)

(new cases 200 to 400 per 100,000).⁸² Before the development of anti-TB drugs in the late 1940s, TB was the leading cause of death in the United States. Drug therapy, along with improvements in public health and general living standards, resulted in a marked decline in incidence. However, recent influxes of immigrants from developing third world nations, rising homeless populations, prolonged lifespans, and the emergence of HIV led to an increase in reported cases in the mid 1980s, reversing a 40-year period of decline.

Overall, between 1985 and 1992 there was a 20% increase in new TB cases in the United States. Now, after years of rising TB infection rates, the United States has started to see a decrease in the annual number of cases (current incidence is 4.8 cases per 100,000 U.S.-born individuals, which is a 3.8% decline, and the incidence among foreign-born persons has not risen).^{33,221} Cases of multidrug-resistant TB have continued to rise annually, and there remains a huge reservoir of individuals who are infected.

Multidrug-resistant TB has emerged as a major infectious disease problem throughout the world. The infected individual begins taking the prescribed medication, feels better, and discontinues taking the drugs that are normally required to be taken 6 to 9 months. The disease flares up months later and is now resistant to the medications, and the infected person passes it along as a new drug-resistant strain characterized by mutations in existing genes.¹³⁹

The AIDS pandemic, the increased incidence of TB in populations without easy access to anti-TB medications (e.g., homeless people and economically disadvantaged people), the deterioration of the public health infrastructure, interruptions in the drug supply, and inadequate training of health care providers in the epidemiology of TB are some factors contributing to the increased incidence of multidrug-resistant TB.

Risk Factors

Although TB can affect anyone, certain segments of the population have an increased risk of contracting the disease, particularly those with HIV infection and people age 65 years and older. The latter constitute nearly one-half of the newly diagnosed cases of TB in the United States and most cases of reactivation of dormant mycobacteria.

Other groups at risk include (1) economically disadvantaged or homeless people living in overcrowded conditions, frequently ethnic groups such as Hispanics, Native Americans, and Asian/Pacific Islanders; (2) immigrants from Southeast Asia, Africa (high HIV incidence), Eastern Europe, Mexico, and Latin America; (3) clients dependent on injection drugs, alcohol, or other drugs associated with malnutrition, debilitation, and poor health; (4) infants and children under the age of 5 years; (5) current or past prison inmates; (6) people with diabetes mellitus; (7) people with end-stage renal disease; and (8) others who are immunocompromised (not only those who are HIV infected but also those who are malnourished, organ transplant recipients, anyone receiving cancer chemotherapy or prolonged corticosteroid therapy).

The Institute of Medicine (IOM) estimates that more than one-half of all TB cases in the United States are attributable to foreign-born residents. The IOM has published a report calling for TB screening of all U.S. immigrants to prevent a predicted resurgence of the disease in the United States.²¹²

Limited access to health care because of socioeconomic status or illegal alien status and sociocultural differences contribute to delays in seeking care and influence adherence to treatment, contributing to the rise in TB among ethnic groups, especially along the U.S.-Mexican border.³⁰⁴

Risk associated with processing contaminated medical waste has been reported,²²⁷ and the first documented case of cadaver-to-embalmer (mortician) TB has occurred, possibly by exposure to infectious aerosols generated during the aspiration of blood and other body fluids from the cadaver.⁴⁰⁰

Staff members of laboratories and necropsy rooms are estimated to be between 100 and 200 times more likely than the general public to develop TB by the inhalation of the bacilli in aerosols or dried material, by injuries (e.g., cuts and accidental inoculations with infected instruments), and by contact with infected materials and surfaces. Necropsies on individuals who had undiagnosed TB while alive present a potential hazard to pathologists, technicians, and medical students involved in postmortem examinations.⁹²

Environmental factors that enhance transmission include contact between susceptible persons and an infectious person in relatively small, enclosed spaces (e.g., evidence of limited transmission during extended airline, train, or bus travel has been documented)²³⁹; inadequate ventilation that results in insufficient dilution or removal of infectious droplet nuclei (e.g., older buildings such as hospitals, prisons, government buildings, universities); and recirculation of air containing infectious droplet nuclei. Adequate ventilation is the most important measure to reduce the infectiousness of the environment. Mycobacteria are susceptible to ultraviolet irradiation (i.e., sunshine), so outdoor transmission of infection rarely occurs.

Etiologic Factors

The causative agent is the tubercle bacillus (Fig. 15-5), commonly transmitted in the United States by inhalation of infected airborne particles, known as droplet nuclei, which are produced when the infected persons sneeze, laugh, speak, sing, or cough.

Casual contact or brief exposure to a few bacilli will not result in transmission of sufficient bacilli to infect a person. Rather, prolonged exposure in an enclosed space is required for transmission. Genetic factors determining susceptibility and resistance to the infection are suspected but have not been proven. In some other parts of the world, bovine TB carried by unpasteurized milk and other dairy products from tuberculous cattle is more prevalent.

The tubercle bacillus is capable of surviving for months in sputum that is not exposed to sunlight. Within the body it becomes encapsulated and can lie dormant for decades and then become reactivated years after an initial

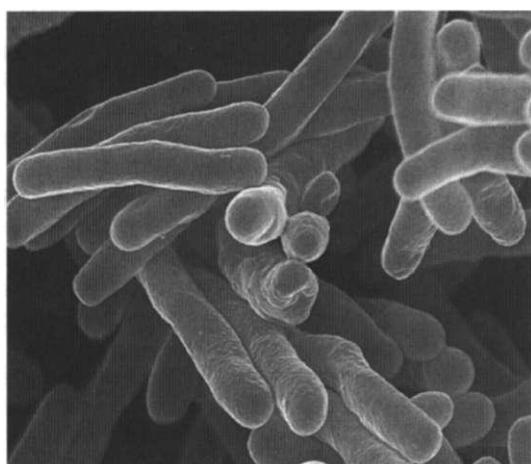


Figure 15-5

Tuberculosis (TB) bacteria. (Courtesy National Institute of Allergy and Infectious Diseases, National Institutes of Health, Bethesda, MD, 2001.)

infection. This secondary TB infection (endogenous reinfestation) can occur at any time the person's resistance is lowered (e.g., alcoholism, immunosuppression, silicosis, advancing age, or cancer).

The older people of today were children when transmission of tubercle bacilli occurred more often. Now, reactivation of the disease is developing in their later years because an increasing portion of older adults who were previously not infected are acquiring new infections in extended care facilities.

Pathogenesis

Once a susceptible person inhales droplet nuclei containing *M. tuberculosis* and bacilli become established in the alveoli of the lungs, a proliferation of epithelial cells surrounds and encapsulates the multiplying bacilli in an attempt to wall off the invading organisms, thus forming a typical tubercle.

Two to ten weeks after initial human infection with the bacilli, acquired cell-mediated immunity usually limits further multiplication and spread of the TB bacilli. Although the TB bacilli are walled off inside a tubercle, the bacilli are not necessarily destroyed; they can remain alive but dormant inside the structure.

No one yet knows how the TB bacterium does its damage. *M. tuberculosis* has no known endotoxins or exotoxins so there is no immediate host response to the infection. The organisms grow for 2 to 12 weeks until they reach a number sufficient to elicit a cellular immune response that can be detected by a reaction to the tuberculin skin test.

The organisms tend to be localized or focused at sites of infection. In persons with intact cell-mediated immunity, collections of activated T cells and macrophages form granulomas that limit multiplication and spread of the organism, rendering the infection inactive, or *latent*. The tubercles stay intact as long as the immune system is maintained.

For the majority of individuals with an intact immune system, latent infection is clinically and radiographically undetected; a positive tuberculin (protein purified deriva-

tive [PPD]) skin test result is the only indication that infection has taken place. Individuals with latent TB infection but not active disease are not infectious and cannot transmit the organism. When residual lesions are visible on chest radiograph these sites remain potential lesions for reactivation.

If, however, the infection is not controlled by the immune defenses, the person develops symptoms of progressive primary TB. The granulomas become necrotic in the center and eventually produce fibrosis and calcification of the tissues.

Tubercle bacilli can spread to other parts of the body by way of the lymphatics to the hilar lymph nodes and then through the bloodstream to more distant sites, producing a condition called *miliary* (evenly distributed small nodules) TB, most common in people 50 years or older and in very young children with unstable or underdeveloped immune systems.

Erosion of blood vessels by the primary lesion can cause a large number of bacilli to enter the circulatory system, where they are carried to all areas of the body and may lodge in any organ, especially the lymph system, spine and weight-bearing joints, urogenital system, and meninges. Untreated, these tiny lesions spread and produce large areas of infection (e.g., TB pneumonia, tubercular meningitis). The same pharmacologic treatment is used for extrapulmonary and pulmonary TB, though the duration may be extended for some neurologic or skeletal infections.¹⁶⁶

Researchers have identified a segment of deoxyribonucleic acid (DNA) that allows the TB organism to invade macrophages, where they lie dormant for years before leaving the macrophage cells to invade the lungs or other parts of the body. Finding this genetic fragment may provide information needed to block the microorganisms from entering human cells. The DNA fingerprint identified probably represents only one of several mechanisms that permit the TB transmission and invasion.¹⁹³ Using DNA, scientists are developing faster and more accurate diagnostic tests for TB. Earlier detection improves treatment effectiveness.²²⁵

Clinical Manifestations

Most symptoms associated with TB do not appear in the early, most curable stage of the disease, although a skin test administered would be positive. Often symptoms are delayed until 1 year or more after initial exposure to the bacilli. Symptoms suggestive of TB include productive cough of more than 3 weeks' duration, especially when accompanied by other symptoms such as weight loss, fever, night sweats, fatigue, malaise, and anorexia. Rales may be heard in the area of lung involvement, as well as bronchial breath sounds, if there is lung consolidation.

Complications associated with TB can include bronchopleural fistulae, esophagopleural fistulae, pleurisy with effusion, tuberculous pneumonia or laryngitis, and sudden lung atelectasis, indicating that a deep tuberculous cavity in the lung has perforated or created an opening into the pleural cavity, allowing air and infected material to flow to it (Fig. 15-6).

Extrapulmonary involvement (e.g., abdominal, peritoneal, genitourinary, lymph node, central nervous

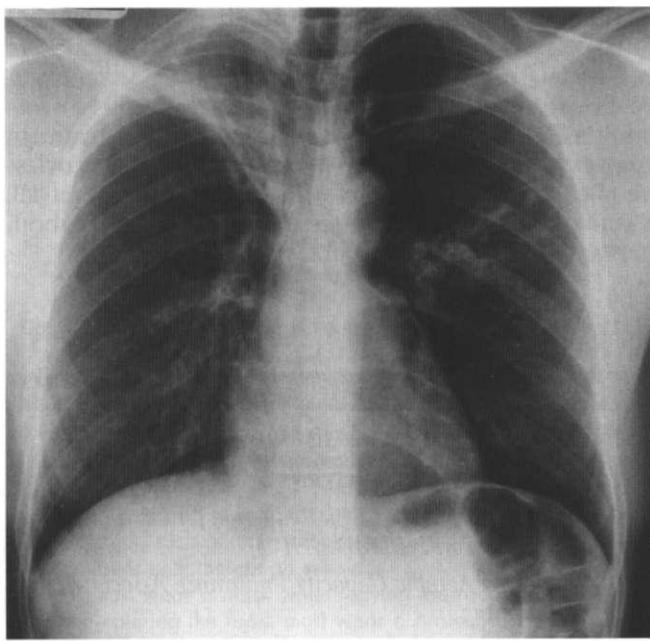


Figure 15-6

Segmental consolidation in tuberculous bronchopneumonia. The right upper lobe is grossly collapsed, scarred, and bronchiectatic. It had remained stable for many years until segmental nodular and linear consolidation appeared in the left mid zone, signaling reactivation. The segmental lesion was thought to be secondary to aspiration of bacteria from the right upper lobe. (From Grainger RG, Allison D: *Grainger and Allison's diagnostic radiology: a textbook of medical imaging*, ed 4, Philadelphia, 2001, Churchill Livingstone. Used with permission.)

system, or skeletal TB) increases in frequency in the presence of declining immunocompetency. Extrapulmonary TB occurs alone (i.e., without pulmonary involvement) in one-third of HIV-infected persons and in another one-third of HIV-infected persons with pulmonary involvement.¹²

Tuberculous involvement of the brain and spinal cord (extrapulmonary TB) is a common neurologic disorder in developing countries and has recently shown resurgence when associated with HIV. In tuberculous meningitis the process is located primarily at the base of the brain, and symptoms include those related to cranial nerve involvement, as well as headache, decreased level of consciousness, and neck stiffness. Tuberculous meningitis is associated with high morbidity and mortality. Tuberculous spondylitis (Pott's disease), a rare complication of extrapulmonary TB, is discussed in Chapter 25.

MEDICAL MANAGEMENT

PREVENTION. Preventing the transmission of TB is essential and can be done by using such simple measures as covering the mouth and nose with a tissue when coughing and sneezing, reducing the number of organisms excreted into the air. However, preventive and therapeutic interventions must address not only the bacillus but also the financial, nutritional, and employment status of those people at risk.

Adequate room ventilation and preventing overcrowding such as in homeless shelters and prisons are well-known preventive measures, but preventing this infection

in many high-risk groups is complicated. For example, should control efforts among the poor emphasize the amelioration of social problems or merely the ingestion of appropriate dose and duration of antibiotic therapy?

Involuntary isolation is no longer acceptable, and directly observed therapy (i.e., the client receives the antibiotics under the supervision of an outreach worker) may be a violation of civil rights. How are individuals' civil liberties and the public health best balanced? How should health professionals address the problem of the noncompliant individual? The complex issues surrounding TB in the United States remain an unresolved challenge at this time.*

The term *preventive drug therapy* has been changed to *treatment of latent TB infection* (LTBI). The failure of vaccination with bacille Calmette-Guerin (BCG), a freeze-dried preparation of a live, attenuated strain of *M. bovis*, to control the global TB epidemic and the spread of multidrug resistance has resulted in renewed research efforts to develop a better vaccine. New vaccines that could boost BCG could be soon available, and live vaccines are currently being tested.²⁷⁹

DIAGNOSIS. Recent advances in DNA molecular techniques for the diagnosis of TB have improved the accuracy and speed of laboratory diagnosis in symptomatic people. Fortunately, some of these improved tools are appropriate for low-income settings and may help integrate new diagnostic tools into national TB control

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programs.¹

Diagnostic measures for identifying TB currently include history, physical examination, tuberculin skin test, chest radiograph, and microscopic examination and culture of sputum. The tuberculin skin test determines whether the body's immune response has been activated by the presence of the bacillus. The skin and other tissue become sensitized to the protein part of the tubercle bacilli. A positive reaction causes a swelling or hardness at the site of infection and develops 3 to 10 weeks after the initial infection. A positive skin test reaction indicates the presence of a TB infection but does not show whether the infection is dormant or is causing a clinical illness. Other diagnostic methods, such as sputum analysis, bronchoscopy, or biopsy, may be indicated in some cases.

Because of the dormant properties of the tubercle bacillus, anyone infected with TB should have periodic TB testing performed. In the case of someone with known TB, the skin test will always be positive, requiring periodic screening with chest x-ray studies. Previously an annual examination was recommended, but currently, screening is based on symptomatic presentation (if asymptomatic, testing is not required) and job exposure (i.e., those health care workers treating persons with active TB or AIDS or HIV infection are at increased risk of exposure).

*For an interesting and insightful look at these and other issues related to TB, the reader is referred to Lerner BH: *Contagion and confinement: controlling tuberculosis along the skid row*, Baltimore, 1998, Johns Hopkins University Press.

The tuberculin skin test is the most common method currently used that demonstrates infection with *M. tuberculosis* in the absence of active TB, although newer methods, including a blood test, QuantiFeron-TB Gold test (QFT-G), have been approved by the FDA. The test detects the release of interferon- γ (IFN- γ) in fresh heparinized whole blood from sensitized persons when it is incubated with two synthetic peptides that simulate two proteins present in *M. tuberculosis*.^{62,225,331}

The new test has higher sensitivity and specificity (fewer false negative and false positive tests) than the old TB PPD skin test and the former QFT test, which used PPD as the incubating agent. The peptide-sensitizing agents used in the test are absent from all BCG vaccine strains and most commonly encountered non-TB mycobacteria. The client only needs one appointment for the test. Nucleic acid amplification techniques are under development.^{225,331}

TREATMENT. The American Thoracic Society and the CDC¹³ have published guidelines for the treatment of TB infection. These guidelines should contribute to improved TB control worldwide and to TB elimination in the United States. Once diagnosed, all cases of active disease are treated, and certain cases of inactive disease are treated prophylactically, although it is unclear that preventive treatment is helpful in people with latent TB.^{13,8} Treatment may be initiated with only a positive skin test even if chest film and sputum analyses show no evidence of the disease. In this way, the disease is less likely to reactivate later in life when the immune system is more likely to be compromised.

Pharmacologic treatment through medication is the primary treatment of choice and renders the infection noncontagious and nonsymptomatic. These agents work by inhibiting cell wall biosynthesis, but the intracellular response that occurs is complex and poorly understood at this time.

Drug treatments now include combinations of all primary anti-TB medications (e.g., rifampin; isoniazid; pyrazinamide [Rifater]; and ethambutol) taken in one dose to replace the traditional treatment requiring multiple drugs daily. Treatment is problematic with homeless people and people who abuse alcohol and use injection drugs because this population is often noncompliant with the recommended 6- to 9-month treatment regimen. Children are usually treated with isoniazid and rifampin for 6 months. Multidrug-resistant TB has further complicated treatment. Treatment of resistant mycobacteria or the complications of TB frequently requires pneumonectomy.

Chemotherapy using a variety of chemical agents may be used, and often two or more drugs are used simultaneously to prevent the emergence of drug-resistant mutants. Immune amplifiers, such as IFN- γ , IL-2, and IL-12, are being tested as possible treatment alternatives. Treatment regimens do not differ for pulmonary and extrapulmonary TB.

PROGNOSIS. Pulmonary TB is a major cause of morbidity and mortality worldwide, resulting in the greatest number of deaths from any one single infectious agent. This trend

is due in part to increasing numbers of individuals infected with both HIV and TB. Untreated, TB is 50% to 80% fatal, and the median time period to death is 2½ years. HIV-related death from TB represents 12% of all adult AIDS-related deaths.⁹³ Noncompletion of treatment (especially among inner city residents and homeless people) is the primary factor in multidrug-resistant TB. Mortality from multidrug-resistant TB is high in both persons infected with HIV and persons free of HIV.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-5

Pulmonary Tuberculosis

PREFERRED PRACTICE PATTERNS

6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders (AIDS/HIV)

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Various other preferred practice patterns may be appropriate in the case of extrapulmonary TB, depending on the associated complications: 6H for lymph node involvement; 7C, 7D, and 7E for integumentary (skin) involvement; 4A, 4B, 4G, and other patterns for musculoskeletal involvement; 5F for cranial nerve involvement; or 5C and 5D for meningitis.

Health care workers should be alert at all times to the need for preventing TB transmission when cough-inducing procedures are being performed, but especially in cases of known TB or HIV infection. Isolation measures for anyone who may be dispersing *M. tuberculosis* must be taken both in the acute care setting and in outpatient areas. Inpatient rooms must be posted with airborne/acid-fast bacilli (AFB) precautions.

If there is a high degree of suspicion or proved TB, clients should be cared for in negative-pressure isolation rooms while undergoing assessment and/or treatment. Procedures that may generate infectious aerosols should be carried out in similarly ventilated rooms. Precautions must be followed by all health care personnel having contact with clients diagnosed with TB (Box 15-4; see also Appendix A).

Therapists may be asked to assist individuals with a weak cough to generate a stronger one, either to improve ventilation or sometimes to obtain a sputum sample without undergoing the more invasive bronchoscopy. In such cases, the therapist should always check to see if the person has ever been diagnosed with pulmonary TB or had a recent TB test. When in doubt, the therapist should practice self-protective measures such as wearing a high efficiency particulate air (HEPA) respirator or a protective mask. Training in the use of the mask and the proper sizing for the therapist are important when using these devices.

Box 15-4**GUIDELINES FOR THERAPISTS FOR PREVENTING TRANSMISSION OF TUBERCULOSIS (TB)**

All TB control recommendations for inpatient facilities apply to hospices and home health services and outpatient settings. All facilities should have a supervisor in charge of infection control compliance.

All new employees (and student therapists) should be screened with the two-step tuberculin skin test or blood assay *M. tuberculosis* test (BAMT).

Doors to airborne infection isolation (AI) rooms must be kept closed.

Clients infected with TB must cover mouth and nose with tissues when coughing, sneezing, or laughing.

Cough-inducing procedures should not be performed on TB clients unless absolutely necessary; such procedures should be performed using local exhaust, in a HEPA-filtered booth or individual TB isolation room. After completion of treatment, such persons should remain in the booth or enclosure until the cough subsides.

Clients must wear a mask when leaving the room.

Anyone entering the room must wear a protective mask, or HEPA respirator, properly.

Therapists must be adequately trained in the use and disposal of masks and should use a PR, which is a special mask,* whenever the client is undergoing cough-inducement or aerosol-generating procedures.

The therapist must check the condition of both the face piece and face seal each time the PR is worn.

Gloves are worn when touching infective material.

Disinfect the stethoscope between treatment sessions.

Staff and employees attending clients in all settings must be tested for TB: every 6 months for high-risk therapists and other personnel annually.

Handwashing is required before and after contact with the client.

Isolation precautions must be continued until a clinical and bacteriologic response to medical treatment has been demonstrated.

Environmental surfaces (e.g., walls, crutches, bed rails, and walkers) are not associated with transmission of infections; only routine cleaning of such items is required.

Therapists with current pulmonary or laryngeal TB should be excluded from work until adequate treatment is instituted, cough is resolved, and sputum is free of bacilli on three consecutive smears.

Home health personnel can reinforce client education about the importance of taking medications as prescribed.

From Guidelines for preventing the transmission of *M. tuberculosis* in health-care facilities, MMWR 54(17):1-141, 2005.

HEPA, High-efficiency particulate air; PR, particulate respirator.

*There are several types of face masks designated as particulate respirators; all National Institute for Occupational Safety and Health (NIOSH)-certified respirators are acceptable protection for health care workers against *M. tuberculosis*. The respiratory protection standard set by the Occupational Safety and Health Administration (OSHA) requires a NIOSH-certified respirator; when such a respirator is used, the law requires that a training and fit-test program be present.

For the therapist evaluating a client with pulmonary TB, a thorough chest assessment and musculoskeletal evaluation should be performed. Chest expansion may be decreased because of diffuse fibrotic changes in progressive disease. Tracheal deviation may be present if there is a significant loss of volume in the upper lobes. Postural adaptations may have developed in late stages of the disease because of poor breathing patterns.¹⁴⁷ Other areas of assessment should include overall posture, gait, muscle strength, balance, and functional mobility. The therapist is referred to the *Guide to Physical Therapist Practice* for examination guidelines, as well as considerations for evaluation and plan of care.

People with TB typically have a poor nutritional status and a progressive weight loss that may have secondary effects on the musculoskeletal system such as postural defects and trigger point irritability. The effects of isolation result in disuse atrophy and cardio-pulmonary and physical deconditioning, including progressive dyspnea.

Finding the balance between exercise and clinical limitations is challenging and there is little evidence that exercise is effective in people with active TB. Specific guidelines for evaluation of medical status during exercise are offered by Galantino and Bishop.¹⁴⁷ Exercise training for patients with post-TB lung disorders has been shown to be effective in improving oxygen uptake, dyspnea, functional outcomes, and timed walking distance.^{14,460}

Side effects of the medication can lead to peripheral neuritis that may be brought to the attention of the therapist. This and any other complication, such as hepatitis, hemoptysis, optic neuritis, or purpura, should be reported to the physician.

Extrapulmonary TB is much less common than pulmonary TB but occurs in 50% of individuals with concurrent HIV. Musculoskeletal and nervous system lesions are prevalent in extrapulmonary TB cases. Treatment of Pott's disease follows the same chemotherapy regimen, with prompt response. Immobilization and avoidance of weight bearing may be required to relieve pain, with attention to maintaining strength and range of motion (see further discussion in Chapter 25).

For the Health Care Worker

For the health care worker who is exposed to TB and develops active disease, treatment will yield a "cure" if the appropriate pharmacologic intervention is followed for the full course prescribed, usually a minimum of 6 months. A cure simply means the active TB will not likely recur, but the person can be reexposed and reinfected. Treatment failure (not taking enough medication or for long enough duration) is a more likely outcome than reinfection/redisease because treatment compliance (i.e., noncompliance) is a much bigger problem.

A person with active disease who misses 2 weeks of treatment must restart the entire course and risks the development of drug-resistant infection. After 2 weeks

Continued.

on effective medication, more than 85% of people with positive sputum cultures convert to a noninfectious status. Although the individual is no longer considered infectious, a minimum of 6 months is required before the disease is considered cured.

If the health care worker is exposed and infected but does not develop active disease (approximately 90% of all cases), there is a 10% lifetime risk that active disease can develop; one-half of that risk takes place in the first 2 years. That 10% risk can be reduced to approximately a 1% risk if a single prophylactic medication is taken properly for 6 months.

In such cases, the individual is considered a "TB reactor" and will always skin test positive for TB. These individuals will require TB clearance in order to work in health care settings, schools, or other similar settings. Clearance is provided via medical documentation of treatment and with a letter from the attending physician.

The TB bacterium must be inhaled and cannot be transmitted by physical contact with extrapulmonary sites unless the organism is expelled, aerosolized, and then inhaled. Although unusual, this type of situation may be encountered during wound care involving the integument and should be approached with appropriate standard precautions.

Lung Abscess

Definition

Described as a localized accumulation of purulent exudate within the lung, an abscess usually develops as a complication of pneumonia, especially aspiration and staphylococcal pneumonia. This can occur when bacteria are aspirated from the oropharynx along with foreign material or vomitus, or it can occur from septic embolus from a heart valve. Septic pulmonary emboli from staphylococcal endocarditis of the tricuspid or pulmonary valves are most often a complication of the use of illicit injection drugs. An abscess may also form when a neoplasm becomes necrotic and contains purulent material that does not drain from the area because of partial or complete obstruction.

Risk Factors

Aspiration associated with alcoholism is the single most common condition predisposing to lung abscess. Other predisposed persons include those with altered levels of consciousness because of drug or alcohol use as mentioned, seizures, general anesthesia, lung cancer, or CNS disease; impaired gag reflex as a result of esophageal disease or neurologic disorders; poor dentition and periodontal care; and tracheal or nasogastric tubes, which disrupt the mechanical defenses of the airways.

Pathogenesis and Clinical Manifestations

As with all abscesses, a lung abscess is a natural defense mechanism in which the body attempts to localize an

infection and wall off the microorganisms so these cannot spread throughout the body. As the microorganisms destroy the local parenchymal tissue (including alveoli, airways, and blood vessels), an inflammatory process causes alveoli to fill with fluid, pus, and microorganisms (consolidation). Death and decay of consolidated tissue may progress proximally until the abscess drains into the bronchus, spreading the infection to other parts of the lung and forming cavities (cavitation).

Clinical signs and symptoms of abscess formation almost always include cough productive of foul-smelling sputum and persistent fever. Other characteristic features include chills, dyspnea, pleuritic chest pain, cyanosis, and clubbing of fingernails, which can develop over a short period of time. Cavitation causes severe cough with copious amounts of purulent sputum and sometimes hemoptysis.

MEDICAL MANAGEMENT

DIAGNOSIS. The radiographic appearance of a thick-walled solitary cavity surrounded by consolidation suggests lung abscess but must be differentiated from other possible lesions. Cavitary lesions in the apex of the upper lobes are frequently caused by TB rather than bacterial abscess. Sputum analysis and culture, bronchoscopy, or ultrasound-guided transthoracic needle biopsy may be diagnostic; the latter diagnostic procedure also permits successful drainage of pulmonary abscesses.

TREATMENT AND PROGNOSIS. Treatment includes specific antibiotics and good nutrition. Airway clearance techniques may be helpful if the abscess communicates with the main-stem bronchi; percussion helps promote drainage of associated secretions. Other measures are similar to the treatment of pneumonia. Bronchoscopy may be used to drain the abscess. Percutaneous drainage has also been deemed to be effective and safe for refractory lung abscess.¹³⁷

Prognosis is good if antibiotics can treat the underlying cause, leaving only a residual lung scar. However, mortality remains in the range of 5% to 10% and is influenced by the severity of the primary disease that initially caused consolidation, the client's general state of health, and the promptness of treatment.¹³⁷

Pneumonitis

Pneumonitis, an acute inflammation of lung tissue usually caused by infections, is discussed in this chapter (see section in this chapter on Environmental and Occupational Diseases) under its most common presentation as hypersensitivity pneumonitis. Other causes of pneumonitis include lupus pneumonitis associated with systemic lupus erythematosus (SLE), aspiration pneumonitis associated with inspiration of acidic gastric fluid, obstructive pneumonitis associated with lung cancer, and interstitial pneumonitis associated with AIDS. Consolidation with impaired gas exchange may occur in the involved lung tissue, but with successful inactivation of the infecting agent, resolution occurs with restoration of normal lung structure.

Acute Bronchitis

Acute bronchitis is an inflammation of the trachea and bronchi (tracheobronchial tree) that is of short duration (1 to 3 weeks) and self-limiting with few pulmonary signs. It may result from chemical irritation, such as smoke, fumes, or gas, or it may occur with viral infections such as influenza, measles, chickenpox, or whooping cough. These predisposing conditions may become apparent during the therapist's interview with the client.

Symptoms of acute bronchitis include the early symptoms of an URI or a common cold, which progress to fever; a dry, irritating cough caused by transient hyperresponsiveness; sore throat; possible laryngitis; and chest pain from the effort of coughing. Later, the cough becomes more productive of purulent sputum, followed by wheezing. There may be constitutional symptoms, including moderate fever with accompanying chills, back pain, muscle pain and soreness, and headache.

Clients with viral bronchitis present with a nonproductive cough that frequently occurs in paroxysms and is aggravated by cold, dry, or dusty air. Bacterial bronchitis (common in clients with COPD) causes retrosternal (behind the sternum) pain that is aggravated by coughing.

Acute bronchitis should be differentiated from chronic bronchitis, pneumonia, whooping cough, rhinosinus conditions, and gastrointestinal reflux disease before treatment begins.⁵⁷ Treatment is conservative and symptomatic with cough suppressants, rest, humidity, and nutrition and hydration.

Seasonal vaccination of people with recurrent bouts of bronchitis reduces the number and severity of exacerbations over the winter months.¹³⁷ Bronchodilators are not indicated and the use of antibiotics for acute bronchitis is not recommended.^{57,442}

Prognosis is usually good with treatment, and although acute bronchitis is usually mild, it can become complicated in people with chronic lung or heart disease and in older adults because they are more susceptible to secondary infections. Pneumonia is a critical complication.

OBSTRUCTIVE DISEASES

Chronic Obstructive Pulmonary Disease

Definition

COPD, also called *chronic obstructive lung disease*, refers to chronic airflow limitation that is not fully reversible. Chronic bronchitis, obstructive bronchiolitis,⁴¹¹ and emphysema are three forms of pathology that manifest as COPD. Chronic, unremitting asthma may be indistinguishable from COPD.⁷⁹ Although these diseases share a common obstructive component and can occur independently, they most commonly coexist, requiring differing treatment and having different prognoses. COPD has also been divided into septic (bronchiectasis) and nonseptic (emphysema) categories to help guide treatment.¹¹⁸

Incidence and Risk Factors

COPD is second only to heart disease as a cause of disability in adults under 65 years of age. It is the fourth leading cause of death in the United States, predicted to be the third leading cause of death by 2020. Nearly 12 million people in the United States were diagnosed with COPD in 2000, but the prevalence is much higher (nearly 24 million adults have documented lung impairment). There were approximately 119,000 deaths from COPD in 2000. The estimated cost of COPD was \$32.1 billion in 2002.

COPD is almost always caused by exposure to environmental irritants, especially smoking, which is the most common cause of COPD; this condition rarely occurs in nonsmokers. As with all chronic diseases, the prevalence of COPD is strongly associated with age and usually presents at age 55 to 60 years. More men are affected than women, but the incidence in women is increasing with the concomitant increase in smoking by women. Because smoking is the major cause of both emphysema and chronic bronchitis, these two conditions often occur together.

Morbidity and mortality rates for COPD increase with the effects of repeated or chronic exposure to irritating gases, dusts, or allergens; chronic irritation; and pollution in urban environments. Other contributing factors include chronic respiratory infections (e.g., sinusitis), periodontal disease,³⁷⁴ the aging process, heredity, and genetic predisposition.

Pathogenesis and Clinical Manifestations

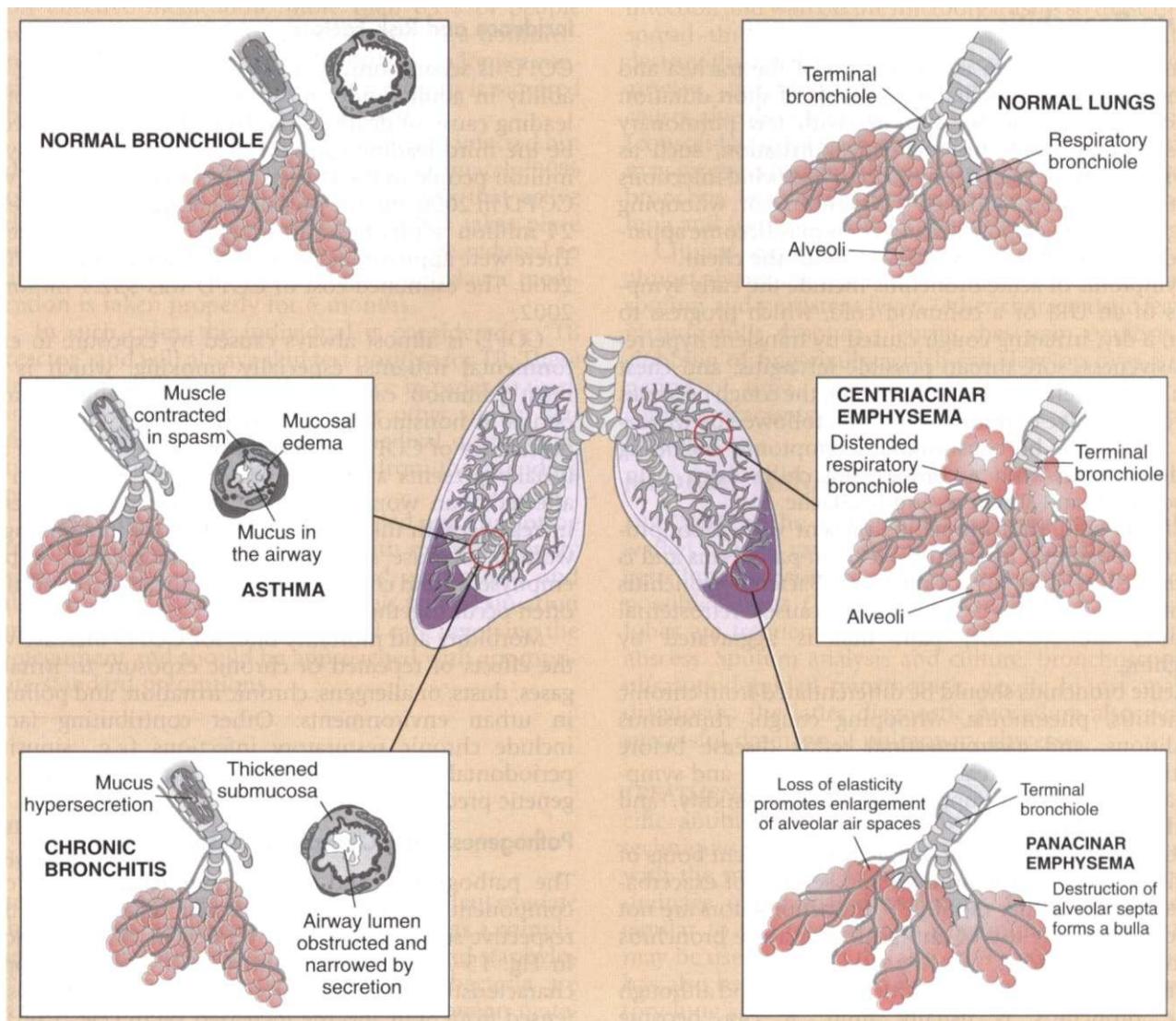
The pathogenesis and clinical manifestations of each component of COPD are discussed separately in their respective sections. A broad overview of COPD is shown in Fig. 15-7. The person with COPD often develops a characteristic look with shoulders raised and muscles tensed from SOB and the increased WOB (Fig. 15-8).

MEDICAL MANAGEMENT

At least two sets of guidelines for diagnosis and management of COPD are available.¹⁶⁵ One set of guidelines was developed by the Global Initiative for Chronic Obstructive Lung Disease (GOLD), a joint project of the National Heart, Lung, and Blood Institute and the World Health Organization. Guidelines were also developed jointly by the American Thoracic Society and European Respiratory Society. These groups have set standards and made recommendations for management and future research of COPD.

DIAGNOSIS. Physical examination and air-flow limitation on pulmonary function testing (see Table 40-22) are assessment tools in determining the presence and extent of COPD. A simple and inexpensive portable spirometer permits such testing in the outpatient setting but may also be conducted in a respiratory laboratory with a computerized spirometer; the results should not be used interchangeably.

Spirometry is the most basic and frequently performed test of pulmonary (lung) function. The spirometer mea-

**Figure 15-7**

What happens in chronic obstructive lung disease/chronic air-flow limitation (COPD).

asures how much air the lungs can hold and how well the respiratory system is able to move air into and out of the lungs. Because spirometry is based on a maximal forced exhalation, the accuracy of its results are highly dependent on the person's understanding, cooperation, and best efforts. Spirometry differs from peak flow readings in that spirometry records the entire forced breathing capacity against time, and peak flow records the largest breathing flow that can be sustained for 10 ms. Both are often used to assess results in the management of asthma.

The spirometer measures the expired air-flow rate and volume in a specific time period. More than a 10% difference measured before and after activity or before and after medication (a bronchodilator) is considered diagnostic for a reactive airway disease component of COPD. Predicted values based on age, height, gender, and body weight are compared to actual values to determine the numeric (%) comparison (Table 15-5).⁷⁷

Spirometry results are expressed as a percentage and are considered abnormal if they are less than 80% of the normal predicted value. An abnormal result usually indicates the presence of some degree of obstructive lung disease. FEV in 1 second (FEV₁) values (percentage of predicted) can be used to classify the obstruction that may occur as mild to very severe.

History, clinical examination, x-ray studies, and laboratory findings usually enable the physician to distinguish COPD from other obstructive pulmonary disorders, such as bronchiectasis, adult CF, and central airway obstruction. High-resolution computed tomography (CT) scan is used to diagnose and quantify emphysema. Most cases of emphysema involve a history of cigarette smoking, chronic cough and sputum production, and dyspnea.

Laboratory analysis may include blood gas measurements and blood pH to indicate the presence of hypoxemia or hypercapnia (excess carbon dioxide in blood)

**Figure 15-8**

Characteristic look of chronic obstructive pulmonary disease (COPD) with shoulders raised and muscles tensed from shortness of breath (SOB) and the increased work of breathing (WOB). This gentleman had a 30-year history of smoking 1.5 packs/day combined with asthma eventually leading to stage IV emphysema and COPD. The effects of asthma and emphysema weakened the heart, resulting in congestive heart failure. Symptoms of SOB, productive cough, fatigue, dizziness, and muscular pain (caused by lack of oxygen) result in disability and reduced quality of life. Use of portable oxygen is required at all times. [Courtesy William T. Cannon, Missoula, MT. Used with permission.]

Table 15-5 Spirometric Classification of Chronic Obstructive Pulmonary Disease (COPD)*

Severity	Postbronchodilator FEV ₁ /FVC	FEV ₁ , %
At risk	>0.7	≥80 predicted
Mild COPD	≤0.7	≥80 predicted
Moderate COPD	≤0.7	50-80 predicted
Severe COPD	≤0.7	30-50 predicted
Very severe COPD	≤0.7	<30 predicted

*The diagnosis of COPD should be considered in anyone who has chronic cough, sputum production, dyspnea, or history of risk factors for COPD. Spirometry is required in making the diagnosis: airflow limitation that is not reversible is indicated by a postbronchodilator FEV₁/FVC ratio of ≤0.7.

FEV₁, forced expiratory volume in 1 second (volume of air expelled in the first second of forced expiration); FVC, forced vital capacity (the maximum volume of air that can be forcibly and rapidly exhaled).

Data from Celli BR, MacNee W: STS/ERS Task Force Standards for the diagnosis and treatment of patients with COPD: a summary of the ATS/ERS position paper, *Eur Respir J* 23(6):932-946, 2004.

and acid-base balance, sputum culture, or presence of immunoglobulin E (IgE) antibodies against specific allergens. Skin testing for allergens that trigger attacks is most useful in young clients with extrinsic allergic asthma.

TREATMENT. The successful management of COPD requires a multifaceted approach that includes smoking

Table 15-6 Pharmacotherapy for Chronic Obstructive Pulmonary Disease (COPD)

Emphysema, Chronic Bronchitis	Asthma
Maintenance	
Inhaled bronchodilators	Antiinflammatory agents
β ₂ -adrenergic agonists (short-acting)	Inhaled corticosteroids
Anticholinergic (antagonize bronchial secretions)	Mast cell stabilizers
Methylxanthines	
Mucolytic expectorants	Bronchodilators
	β ₂ -adrenergic agonists (long-acting)
	Leukotriene antagonists
	Anticholinergics (long-acting)
	Methylxanthines
Acute exacerbations	
Antiinflammatory agents	Inhaled bronchodilators
Oral corticosteroids	β ₂ -adrenergic agonists (short-acting)
Antibiotics	Anticholinergic

Courtesy Susan Queen, PT, PhD, University of New Mexico, Albuquerque, NM.

cessation, pharmacologic management, airway clearance as needed, exercise (aerobic, strength, flexibility, posture, and breathing), control of complications, avoiding irritants, psychologic support, and dietary management.

The main goals for the client with COPD are to improve oxygenation and decrease carbon dioxide retention. These are accomplished by (1) reducing airway edema secondary to inflammation and bronchospasm (asthma) through the use of bronchodilator medication, (2) facilitating the elimination of bronchial secretions, (3) preventing and treating respiratory infection, (4) increasing exercise tolerance, (5) controlling complications, (6) avoiding airway irritants and allergens, (7) relieving anxiety and treating depression, which often accompany COPD, and (8) exercise to improve muscle oxidative capacity.

Common classifications of medications used in the treatment of COPD include oral or inhaled bronchodilators, antiinflammatory agents, antibiotics, mucolytic expectorants, mast cell membrane stabilizers, and antihistamines (Table 15-6). Combining bronchodilators improves effectiveness in reducing exacerbations and improves lung function.¹²³ Systemic corticosteroids are of some help in acute exacerbations of COPD but have some side effects, including hyperglycemia,⁴⁵⁴ and do not produce any long-term benefits.³²⁰

The benefits of pneumococcal vaccine have been proved (decreased mortality and hospitalization), and vaccination is recommended for all people with COPD. Annual prophylactic vaccination against influenza is also recommended.

Narcotics, tranquilizers, and sedatives are used with caution because these depress the respiratory center. Pharmacologic research includes investigation of multiple mediator antagonists, antiinflammatories with better delivery of the medication and lower side effects, induced repair of alveolar tissue, and effects of drug combinations.^{131,123}

Long-term oxygen treatment (LTOT) reduces morbidity and extends life in clients with hypoxemia.⁷ People with Pao_2 of 55 to 59 mm Hg or less (determined by arterial blood gases [ABGs]) with signs of tissue hypoxemia (see Table 15-2) are considered for long-term oxygen therapy. Oxygen therapy is also considered for those who desaturate during sleep or exercise. The National Heart, Lung, and Blood Institute in collaboration with the Centers for Medicare and Medicaid Services has identified areas of future research to improve care and/or reduce cost of care.⁹⁹

Surgical treatment for COPD remains controversial, but lung-volume reduction surgery (LVRS), which is bilateral pneumectomy or removal of large bullae that compress the lung and add to dead space, has been shown to reduce the lung volume, relieve thoracic distention, and improve respiratory mechanics and reduce morbidity.^{78,199} LVRS may be an alternative treatment to lung transplantation for selected individuals with end-stage disease.

A multicenter, prospective, randomized study, called the *National Emphysema Treatment Trial*, was established by the National Heart, Lung, and Blood Institute to study the medical management, including pulmonary rehabilitation, in COPD. Confirming previous research, no change in lung function occurred with rehabilitation, but significant changes were seen in exercise tolerance, dyspnea, and quality of life. Pulmonary rehabilitation is considered important in selecting surgical candidates and preparing them for surgery.³⁵⁵

Lung transplantation, both single and double, are appropriate for clients with COPD when FEV₁ is less than 24% predicted and/or the partial pressure of arterial carbon dioxide (PaCO_2) is equal to or greater than 55 mm Hg. Survival rates are approximately 80% at year 1, 50% at year 5, and 35% at year 10.^{69,281}

PROGNOSIS. The prognosis for chronic bronchitis and emphysema is poor because these are chronic, progressive, and debilitating diseases. The death rate from COPD has increased 22% in the last decade, especially among older men, and the mortality rate 10 years after diagnosis is greater than 50%. COPD is largely preventable, and many believe that early recognition of small airway obstruction with appropriate treatment and cessation of smoking may prevent relentless progression of this disease. Early treatment of airway infections and vaccination against influenza and pneumococcal disease have an effect on morbidity and mortality of individuals with COPD.

There is no cure for COPD, but smoking cessation and oxygen therapy have been shown to increase the survival rate. Pulmonary rehabilitation has also been shown to improve quality of life, decrease hospitalizations, and decrease incidence of COPD exacerbations.^{53,382}

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-6

Chronic Obstructive Pulmonary Disease

PREFERRED PRACTICE PATTERNS

- 4C Impaired Muscle Performance
- 6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders (tobacco disorder)
- 6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning
- 6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction
- 6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure
- 6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Pulmonary Rehabilitation

The pulmonary rehabilitation program model contains many intervention components beyond just airway clearance techniques and exercise, including (but not limited to) smoking cessation, nutrition and weight control, psychosocial support, and lifestyle modification, as well as optimal medical care.¹⁴⁰ Treatment of COPD includes breathing exercises, postural drainage, physical training, a program to improve posture, and strengthening of respiratory musculature. For the motivated child with asthma, breathing exercises and controlled breathing are of value in preventing overinflation, improving the strength of respiratory muscles and the efficiency of the cough, and reducing the WOB.

People with COPD often adopt a sedentary lifestyle, which leads to progressive deconditioning. Deconditioning will lead to progressive deterioration in limb and respiratory muscle function that could adversely affect exercise capacity. The cost and benefits of pulmonary rehabilitation for people with COPD have been well documented.

Although there is little evidence that rehabilitation efforts can result in improved pulmonary function or ABGs, there is strong and growing evidence that programs that include exercise retraining can result in significant benefits such as reduced hospitalizations, increased exercise tolerance, reduced dyspnea, improved skills in using inspiratory muscle training devices, increased independent activities of daily living (ADL) skills, and increased sense of well-being and quality of life.³⁵⁵ Additionally, a recent study demonstrated greater gains in function after training than would be predicted by clinical testing, thus outcome assessment should include functional and quality-of-life tools.³⁸ Various measures of health status have been investigated and are available.^{74,143,270,389}

For clients with α_1 -antitrypsin deficiency (AATD) on augmentation therapy, side effects can include headaches, myalgia (muscle pain), arthralgia (joint pain), and reports of low back pain. Decreased exercise tolerance and increased SOB may occur in individuals with severe COPD or heart failure. The therapist can

be an important educator for anyone with AATD by reminding the individual of the importance of annual flu vaccine and pneumonia shot at intervals determined by the physician. Early signs and symptoms of infection must be reported to the physician with immediate treatment to prevent increased neutrophil count (neutrophils inhibit AAT).

Exercise

Exercise limitation is a common and disturbing manifestation of COPD caused by multiple interrelated anatomic and physiologic disturbances. Exercise tolerance can be improved despite the presence of fixed structural abnormalities in the lung. Only a weak correlation between degree of airway obstruction and exercise tolerance has been shown, suggesting that factors other than lung function impairment (e.g., deconditioning and peripheral muscle dysfunction) play a predominant role in limiting exercise capacity in people with COPD. In fact, inspiratory capacity is a more powerful predictor of exercise tolerance than FEV₁ or forced vital capacity (FVC).¹⁹⁸

Muscle weakness in stable COPD does not affect all muscles the same. For example, proximal upper limb muscle strength may be impaired more than distal upper limb muscle strength, peripheral muscle may be limited mainly by endurance capacity, and the diaphragm muscle may be altered structurally (e.g., changes in muscle length and configuration affecting the mechanical force and action)¹⁷³ and limited in strength capacity. These alterations in different muscle types require individual assessment and exercise prescription.^{173,174}

Clients with COPD must be encouraged to remain active, with specific attention directed toward activities they enjoy. Training in pacing and energy conservation (see Box 9-8) allows even those with limited exercise tolerance to increase their daily activities. Exercise testing, individualized to the specific client's needs and goals, is used to determine the person's baseline and for prescribing a training regimen. Medications and oxygen may be used or timed to determine the effect of these interventions. Oximetry, heart rate, respiratory rate, and blood pressure are routinely monitored, and blood gases or expired gases may be used to assess response to testing. Respiratory muscle strength testing is measured as maximum inspiratory and expiratory pressures.¹⁴⁰

Strengthening the muscles of respiration is done by teaching the person with COPD to take slow, deep breaths and the use of resistive devices. Pursed-lip breathing helps slow the respiratory rate and prevent airway collapse during exhalation. The client is instructed to inhale through the nose and exhale with the lips pursed in a whistling or kissing position. Each inhalation should take about 4 seconds and each exhalation about 6 seconds.

Effective coughing techniques using diaphragmatic breathing help to mobilize secretions. Ventilatory muscle training should be included for anyone who continues to experience exercise limitation and breathlessness despite medical therapy and general exercise

reconditioning. Respiratory muscle training improves exercise tolerance, dyspnea, and quality of life.²⁴⁷

People with chronic air-flow obstruction report disabling dyspnea when performing seemingly trivial tasks (e.g., activity with unsupported arms). Some muscles of the upper torso and shoulder girdle share both a respiratory and a positional function for the arms, resulting in functional limitations in many clients with lung disease during unsupported upper extremity activities.

Simple arm elevation results in significant increases in metabolic and ventilatory requirements in clients with long-term air-flow limitations. Pulmonary rehabilitation that includes upper extremity training (progressive resistance exercises [PREs]) reduces metabolic and ventilatory requirements for arm elevation. This type of program may allow clients with COPD to perform sustained upper extremity activities with less dyspnea.¹⁵⁴

Lower extremity training should be included routinely in the exercise prescription. The choice of type and intensity of training should be based primarily on the individual's baseline functional status, symptoms, needs, and long-term goals. Progressively increased walking is the most common form of exercise for COPD. Swimming is a preferred exercise option for clients with bronchial asthma (see next discussion). Therapists working with these clients should encourage them to maintain hydration by drinking fluids (including before, during, and after exercise) to prevent mucous plugs from hardening and to take medications as prescribed. When tolerated, high-intensity (continuous or interval), short-term training may lead to greater improvements in quality of life and aerobic fitness than low-intensity training of longer duration, but it is not absolutely necessary to achieve gains in exercise endurance.^{53,144}

Exercise tolerance may improve after exercise training (including weight training)¹⁹⁹ because of gains in aerobic fitness or peripheral muscle strength,²⁷² enhanced mechanical skill and efficiency of exercise, improvements in respiratory muscle function, breathing pattern, or lung hyperinflation. Exercise improves muscle oxidative capacity and recovery in individuals with COPD.³⁵⁰

Exercise training can also reduce anxiety, fear, and dyspnea previously associated with exercise in the deconditioned person. Exercises for flexibility, posture, and motor control can improve mechanical and kinetic efficiency and thus can reduce oxygen demand during daily activities and exercise.

Gains made in exercise tolerance, peripheral and respiratory muscle strength, and quality of life can last up to 2 years after a limited duration (6- to 12-week) rehabilitation program.^{53,423} The optimal duration for an exercise program remains unknown; a 7-week course provides greater benefits than a 4-week course in terms of improvements in health status. Further studies in this area are needed.¹⁷⁵

Three training sessions per week improve exercise performance and health status, whereas a program

Continued.

consisting of 2 sessions/week for 8 weeks may not be effective in people with moderate COPD.^{3,57} Maximally intense exercise sustained over 45 minutes daily, 5 days/week for 6 weeks has been reported more effective in endurance training than exercise of a moderate intensity.¹⁵⁸ Other considerations for exercise training in chronic lung disease are available.^{2,169,233,447}

Monitoring Vital Signs

Using a pulse oximeter can help the therapist and client observe for a decrease in oxygen saturation before hypoxemia occurs. Oxygen saturation is generally kept at 90% or above by adjusting supplemental oxygen levels, adjusting activity level, and practicing physiologic modulation or physiologic quieting.²¹⁰ Altering physiologic responses using principles of self-induced biofeedback and breathing techniques may be able to help some clients self-regulate oxygen saturation levels (see reference 210 for a description of these techniques). Some people with COPD retain carbon dioxide and have a depressed hypoxic drive requiring low oxygen levels to stimulate the respiratory drive. In such cases the upward adjustment of supplemental oxygen levels must be monitored very carefully; increasing total oxygen administered via nasal canula higher than 1 to 2 L requires careful monitoring of the respiratory system (e.g., respiratory rate and breathing pattern), documentation, and consultation with other members of the pulmonary rehabilitation team.

Blood pressure and pulse should be observed at rest and in response to exercise, especially in anyone with COPD and cardiac arrhythmias. Most people with COPD who have mild arrhythmias at rest do not tend to have increased arrhythmias during exercise. Arrhythmias may disappear with exercise and increased perfusion. See also the section on Exercise and Arrhythmias in Chapter 12.

In people with expanded lung volumes because of air trapping, such as occurs in COPD (especially emphysema), the first heart sound is best heard under the sternal area (put the scope in the client's left epigastric area) rather than the apical or mitral area. The hyperinflation of the lungs causes the heart to elongate, displacing the left ventricle downward and medially.

Lung sounds are also changed because the loss of interstitial elasticity and the presence of interalveolar septa lead to air trapping with increased volume of air in the lungs. Air pockets are poor transmitters of vibrations; thus vocal fremitus (the client whispers "99, 99, 99"), breath sounds, and the whispered and spoken voice are impaired or absent on auscultation.

This absence of the vesicular quality of lung sounds is distinctive and may be heard before radiographic evidence of COPD. On the other hand, when there is fluid in the lung or lungs, consolidation, or collapse (e.g., atelectasis), whispered words are heard perfectly and clearly. This is the earliest sign of atelectasis.

A peak flowmeter, a home monitoring device to measure fast expiratory flow (a reflection of bronchoconstriction), can be used to determine how compromised a client with asthma or reactive airways may be

compared to the normal values for that person. This may be a useful measure in determining response to therapy intervention and documenting measurable outcomes.

Exercise and Medication

The majority of pulmonary medications are used to promote bronchodilation and improve alveolar ventilation and oxygenation and are delivered as an aerosol spray through a device called a *metered-dose inhaler* (MDI). Older adults sometimes have difficulty using an inhaler because of arthritis or other medical problems that impair hand/breath coordination.

Proper technique is important to ensure delivery of the medication to the desired location (Box 15-5).⁵⁴ When medications are properly used, their effects should improve an individual's ability to exercise and more effectively obtain the benefits of training. However, the many side effects of pulmonary medications may interfere with normal adaptations to habitual exercise so that exercise tolerance and conditioning may not occur.⁷⁰

For example, corticosteroids mask or impede the beneficial effects of exercise. Anyone with pulmonary disease taking prolonged corticosteroids may develop steroid myopathy (see Chapter 5) and muscular atrophy not only in the peripheral skeletal muscles but also in the muscle fibers of the diaphragm. Animal studies also suggest that severe undernutrition causes a decrease in muscle energy status, which contributes to diaphragmatic fatigue.²⁴⁵ Recent studies have shown promise for use of anabolic steroids and L-carnitine supplements in combating muscle changes in COPD.^{49,98}

Chronic Bronchitis

Definition and Overview

Chronic bronchitis is clinically defined as a condition of productive cough lasting for at least 3 months (usually the winter months) per year for 2 consecutive years. If obstructive lung disease characterized by a decreased FEV₁/FVC ratio* less than 75% is combined with chronic cough, chronic bronchitis is diagnosed. Initially, only the larger bronchi are involved, but eventually all airways become obstructed, especially during expiration.

Risk Factors and Pathogenesis

Chronic bronchitis is characterized by inflammation and scarring of the bronchial lining. This inflammation obstructs air flow to and from the lungs and increases mucus production. Irritants, such as cigarette smoke, long-term dust inhalation, or air pollution, cause mucus hypersecretion and hypertrophy (increased number and

*FEV₁ is the forced expiratory volume, a measure of the greatest volume of air a person can exhale during forced expiration; the subscript is added to indicate the percentage of the vital capacity that can be expired in 1 second. FVC is forced vital capacity, a measure of the greatest volume of air that can be expelled when a person performs a rapid, forced expiratory maneuver. This usually takes about 5 seconds.

Box 15-5**USE OF A METERED-DOSE INHALER**

The therapist should observe the client self-administer the medication at least one time. Schedule the use of MDI 15 to 20 minutes before exercise to maximize ventilation.

- Shake the unit for 5 to 10 times.
- Exhale to normal expiratory volume while slightly tilting your head back.
- Place the inhaler mouthpiece in the mouth in one of three positions (client preference):
 1. In the mouth with the tongue and teeth out of the way (not recommended for corticosteroid use)
 2. Resting on the lower lip with the mouth wide open
 3. 1 to 1.5 inches in front of a wide-open mouth; a 4- to 6-inch spacer that fits over the end of the inhaler can be used to maintain the correct distance away while avoiding spraying the eyes
- Press the inhaler and inhale slowly and deeply for 3–5 seconds to avoid depositing the medication on the back of the throat and to ensure delivery to the lungs.
- At the end of exhalation, hold the breath for 10 seconds.
- Exhale through pursed lips; when using steroids, rinse mouth before swallowing.
- Wait a few minutes before repeating the procedure if more than one puff is prescribed.
- To determine whether an inhaler is empty, keep a completely empty (marked) inhaler on hand. Place the empty inhaler and the questionable inhaler in a bowl (or sink) of water. If the one in question floats just as high as the empty one, replace the inhaler. Number of doses is written on the label.
- Rinse the inhaler and spacer after each use.

size) of mucus-producing glands in the large bronchi. Epithelial atrophy, changes in squamous cells, and hypertrophy of smooth muscle cells occur.⁴¹¹

The swollen mucous membrane and thick sputum obstruct the airways, causing wheezing and a subsequent cough as the person tries to clear the airways. In addition, impaired ciliary function reduces mucous clearance and increases client susceptibility to infection. Infection results in even more mucus production with bronchial wall inflammation and thickening. As airways collapse, air is trapped in the distal portion of the lung, causing reduced alveolar ventilation, hypoxia, and acidosis. This downward spiral continues since the client now has an abnormal ventilation/perfusion (V/Q) ratio and resultant decreased Pao_2 .

As compensation for the hypoxemia, polycythemia (overproduction of erythrocytes) occurs. Cyanosis results from insufficient arterial oxygenation and peripheral edema from ventricular failure. Pulmonary vascular resistance caused by inflammation and loss of capillary beds will lead to cor pulmonale (right-sided congestive heart failure).

Clinical Manifestations

The symptoms of chronic bronchitis are persistent cough and sputum production (worse in the morning and evening than at midday). The increased secretion from the bronchial mucosa and obstruction of the respiratory passages interfere with the flow of air to and from the

lungs. The result is SOB, prolonged expiration, persistent coughing with expectoration, and recurrent infection. Infection may be accompanied by fever and malaise.

Over time, reduced chest expansion, wheezing, cyanosis, and decreased exercise tolerance develop. In addition, the obstruction present results in decreased alveolar ventilation and increased $Paco_2$. Hypoxemia (deficient oxygenation of the blood) leads to polycythemia (over-production of erythrocytes) and cyanosis. If not reversed, pulmonary hypertension leads to cor pulmonale. Severe disability or death is the final clinical picture.

MEDICAL MANAGEMENT

In stable conditions, reducing irritants and using a combination of bronchodilators is effective. There is no evidence for use of antibiotics, oral corticosteroids, expectorants or postural drainage. In acute exacerbations of chronic bronchitis, antibiotics and oral or IV corticosteroids can be effective.⁴¹² See also Medical Management and Special Implications for the Therapist: Chronic Obstructive Pulmonary Disease in the previous section in this chapter.

Emphysema**Definition and Overview**

Emphysema is defined as a pathologic accumulation of air in tissues, particularly in the lungs, and is found in the lungs of most people with COPD. There are three types of emphysema (see Fig. 15-7). Centrilobular emphysema, the most common type, produces destruction in the bronchioles, usually in the upper lung regions. Inflammation develops in the bronchioles, but usually the alveolar sac (distal to respiratory bronchioles) remains intact.

Panlobular emphysema destroys the air spaces of the entire acinus and most commonly involves the lower lung. These two forms of emphysema, collectively called *centriacinar emphysema*, occur most often in smokers. Paraseptal (panacinar) emphysema destroys the alveoli in the lower lobes of the lungs, resulting in isolated blebs along the lung periphery. Paraseptal emphysema is believed to be the likely cause of spontaneous pneumothorax.

Etiologic Factors

Cigarette smoking is the major etiologic factor in the development of emphysema and has been shown to increase the numbers of alveolar macrophages and neutrophils in the lung,* enhance protease release,⁴¹³ and

*Neutrophils, the most numerous type of leukocytes (white blood cells), increase dramatically in number in response to infection and inflammation. However, neutrophils not only kill invading organisms but also may damage host tissues when there are too many.

⁴¹³Proteases, or proteolytic enzymes, are enzymes that destroy cells and proteins. The airway goblet cells and serous cells of bronchial glands normally secrete a protein called *secretory leukoprotease inhibitor*, which is capable of inhibiting neutrophils. The cellular interactions associated with smoking result in inactivation of protease inhibitors. This results in an imbalance between proteases and antiproteases (in favor of proteases), allowing even more cellular destruction than warranted by the inflammatory process already present.

impair the activity of antiproteases. However, other factors, such as heredity, must determine susceptibility to emphysema because less than 10% to 15% of people who smoke develop clinical evidence of airway obstruction.

In many cases, emphysema occurs as a result of prolonged respiratory difficulties, such as chronic bronchitis that has caused partial obstruction of the smaller divisions of the bronchi. Emphysema can also occur without serious preceding respiratory problems as in the case of a defect in the elastic tissue of the lungs or in older persons whose lungs have lost their natural elasticity.

A number of clients with early onset of COPD have an inherited deficiency of (low levels or absent) AAT, a protective protein. AAT is made in the liver but circulates in the blood to protect the tissues in the body, including the lungs. Epidemiologists report approximately 100,000 people with this disorder; 20 million more may be undiagnosed asymptomatic carriers. AAT deficiency is suspected in smokers who develop emphysema before age 40 years and in nonsmokers who develop emphysema; symptoms may not develop until affected individuals are in their seventies. Progressive liver (cirrhosis) and lung disease (panlobular emphysema) occur when AAT is absent.⁴⁰⁴

Pathogenesis

Emphysema is a disorder in which destruction of elastin protein in the lung that normally maintains the strength of the alveolar walls leads to permanent enlargement of the acini. It is suspected that an interaction of accelerated cellular apoptosis, inflammation, and proteolysis causes the tissue destruction associated with emphysema.⁴¹¹

Eventually the loss of elasticity in the lung tissue causes narrowing or collapse of the bronchioles so that inspired air becomes trapped in the lungs, making breathing difficult, especially during the expiratory phase. Obstruction results from changes in lung tissues, rather than from mucus production and swelling as in chronic bronchitis (which is why steroids are usually not helpful in this condition).

The permanent overdistention of the air spaces with destruction of the walls (septa) between the alveoli is accompanied by partial airway collapse and loss of elastic recoil. Pockets of air form between the alveolar spaces (blebs) and within the lung parenchyma (bullae). This process leads to increased ventilatory dead space, or areas that do not participate in gas or blood exchange (Fig. 15-9).

The WOB is increased as a result of ventilatory drive from hypoxemia and hypercapnia, increased effort during exhalation (normally passive recoil), and flattening of the diaphragm caused by hyperinflation. As the disease progresses, there is increasing dyspnea and pulmonary infection. Pulmonary hypertension develops from capillary loss and vessel intimal thickening, and this eventually leads to cor pulmonale (right-sided congestive heart failure).

In centrilobular emphysema the destruction of the lung is uneven and originates around the airways. The membranous bronchioles are thicker, narrower, and more reactive than in panlobular emphysema. Lung corn-

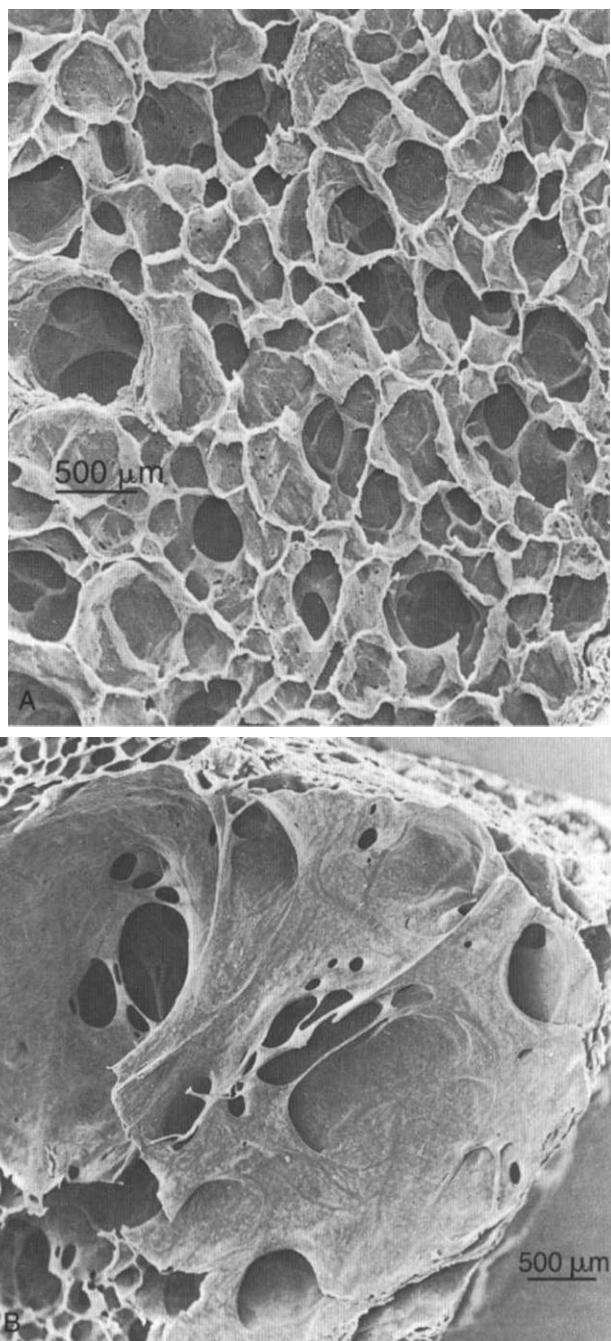


Figure 15-9

Effects of emphysema seen in these scanning electron micrographs of lung tissue. **A**, Normal lung with many small alveoli. **B**, Lung tissue affected by emphysema. Notice that the alveoli have merged into larger air spaces, reducing the surface area for gas exchange. (From Thibodeau GA, Patton KT: *Structure and function of the body*, ed 11, St Louis, 2000, Mosby. Used with permission.)

pliance is low or normal and does not relate to the extent of the emphysema (i.e., not to the losses of elastic recoil), but rather the decrease in airflow is related mainly to the degree of airway abnormality.

In contrast, panlobular emphysema is characterized by even destruction of the lung and the small airways appear less narrowed and less inflamed than in centrilobular

emphysema. Lung compliance is increased and is related to the extent of the emphysema; the decrease in airflow is primarily associated with the loss of elastic recoil rather than with the abnormalities in the airways.⁹⁹

At the molecular and microvascular levels, protease-antiprotease (associated with AATD emphysema) and oxidant-antioxidant theories continue under investigation as theories relate to impaired reparative mechanisms in the causation of emphysema. Oxidative damage by free radicals, which is the basis for the free radical theory of aging (see discussion in Chapter 6), identifies cigarette smoke as the main source of oxidants contributing to epithelial damage associated with smoking-induced emphysema. Determining the mechanisms regulating the antioxidant responses is critical to understanding the role of oxidants in the pathogenesis of smoking-induced lung disease and to developing future strategies for antioxidant therapy.³⁸²

Muscle wasting in emphysema and COPD is linked to increased TNF- α production. Respiratory muscle atrophy can lead to deterioration of lung function and increased WOB.⁴¹¹

Clinical Manifestations

At first, symptoms may be apparent only during physical exertion, but eventually marked exertional dyspnea progresses to dyspnea at rest. This occurs as a result of the irreversible destruction reducing elasticity of the lungs and increasing the effort to exhale trapped air. Cough is uncommon, with little sputum production. The client is often thin, has tachypnea with prolonged expiration, and must use accessory muscles for ventilation. To increase lung capacity and use of accessory muscles, the client often leans forward with arms braced on the knees supporting the shoulders and chest. The combined effects of trapped air and alveolar distention change the size and shape of the client's chest, causing a barrel chest and increased expiratory effort. The normal arterial oxygen levels and dyspnea give clients a classic appearance (Fig. 15-10).

Persons with emphysema have three times the rate of anxiety as the general public. This anxiety is associated with dyspnea or fear of dyspnea. Antianxiety medications, particularly selective serotonin reuptake inhibitors (SSRIs), and cognitive behavioral therapy have been shown to be helpful, although more research is needed.⁶⁰

The most common signs and symptoms of AATD include dyspnea, wheezing, cough, chronic allergies (year round), asthma that does not respond to treatment, and liver problems. A high prevalence of wheezing to allergen and irritant exposures with symptoms of atopy suggests that asthma is common in AATD but is usually associated with COPD. Individuals with AATD who are susceptible to asthma require allergy evaluation and aggressive anti-inflammatory management.¹²⁵

MEDICAL MANAGEMENT

DIAGNOSIS AND TREATMENT. Diagnosis is made on the basis of history (usually cigarette smoking), physical examination, chest film, and pulmonary function tests. The most important factor in the treatment of emphy-

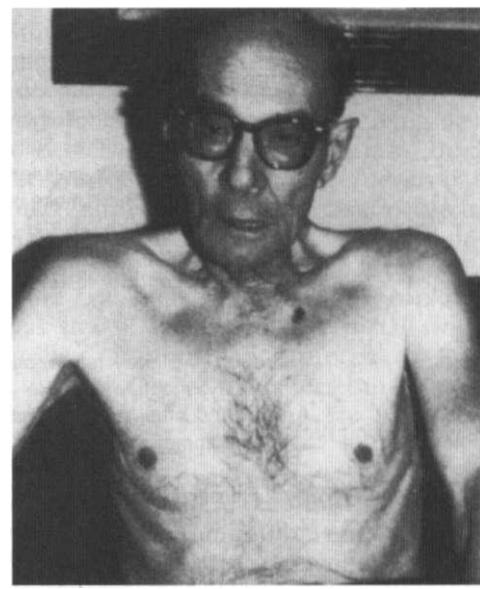


Figure 15-10

The person with emphysema presents with classic findings. Use of respiratory accessory (intercostal, neck, shoulder) muscles and cachectic appearance (wasting caused by ill health) reflect two factors: (1) shortness of breath (SOB), the most disturbing symptom, and (2) the tremendous increased work of breathing (WOB) necessary to increase ventilation and maintain normal arterial blood gases (ABGs). (From Kersten LD: *Comprehensive respiratory nursing*, Philadelphia, 1989, WB Saunders.)

sema is cessation of smoking. Human lungs benefit no matter when someone quits smoking (see Table 3-4); quitting smoking is the most effective way of preventing lung function decline caused by emphysema (and chronic bronchitis).

Pursed-lip breathing causes resistance to outflow at the lips, which in turn maintains intrabronchial pressure and improves the mixture of gases in the lungs. This type of breathing should be encouraged to help the client get rid of the stale air trapped in the lungs. However, this type of breathing pattern has been found to be ineffective for some people; methods to examine diaphragmatic movement and the potential for success with diaphragmatic breathing are available.⁷⁰ Pulmonary rehabilitation and supplemental oxygen are critical aspects of management of COPD.⁷⁸

In the case of AATD, serum testing to measure the levels of AAT is required to identify this problem. Underrecognition of AATD is common with diagnostic and treatment delays documented.⁴⁰³ AATD is treated with weekly intravenous augmentation therapy to slow down or halt the destruction of lung tissue. Home infusion is available for some people.

Infusion of purified AAT from pooled human plasma raises the concentration in serum and epithelial-lining fluid above the protective threshold. Although evidence suggests this treatment slows the decline of lung function and may reduce infection rates while enhancing survival, the cost-effectiveness has been questioned.⁴⁰² Only nonsmokers can benefit from this treatment regimen; smoking increases neutrophils, which in turn inhibit AAT.

Table 15-7 Types of Asthma

Classification	Triggers*
Extrinsic	IgE-mediated external allergens Foods; sulfite additives (wines) Indoor and outdoor pollutants, including ozone, smoke, exhaust Pollen, dust, molds Animal dander, feathers
Intrinsic	Unknown; secondary to respiratory infections
Adult-onset	Unknown
Exercise-induced	Alteration in airway temperature and humidity; mediator release
Aspirin-sensitive (associated with nasal polyps)	Aspirin and other nonsteroidal antiinflammatory drugs
Allergic bronchopulmonary aspergillosis	Hypersensitivity to <i>Aspergillus</i> species
Occupational	Metal salts (platinum, chrome, nickel) Antibiotic powder (penicillin, sulfathiazole, tetracycline) TDI Flour Wood dusts Cotton dust (byssinosis) Animal proteins Smoke inhalation (firefighters) Latex-induced (see Box 4-5)

IgE, Immunoglobulin E; TDI, toluene diisocyanate.

*Emotional stress can be a trigger for anyone with asthma.

PROGNOSIS. Prognosis for individuals with symptomatic AATD is poor with a high incidence of transplantation for liver and lung disease and even more on a transplant waiting list.⁴⁰⁵ Lung volume reduction surgery (surgically removing damaged areas of the lung) may help improve breathing and ventilation. Studies have not been able to predict who can benefit the most from this treatment approach. See also the section in this chapter on Medical Management under Chronic Obstructive Pulmonary Disease and Special Implications for the Therapist: Chronic Obstructive Pulmonary Disease.

Asthma

Definition and Overview

Asthma is defined as a reversible obstructive lung disease characterized by inflammation and increased smooth muscle reaction of the airways to various stimuli. It is a chronic condition with acute exacerbations and characterized as a complex disorder involving biochemical, autonomic, immunologic, infectious, endocrine, and psychologic factors in varying degrees in different individuals. This condition can be divided into two main types according to causative factors: extrinsic (allergic) and intrinsic (nonallergic), but other recognized categories include adult-onset, exercise-induced, aspirin-sensitive, *Aspergillus*-hypersensitive, and occupational asthma (Table 15-7).

Incidence

Asthma, as one component of COPD, is the most common chronic disease in adults (11.9% incidence) and children (7% to 9% incidence), and the prevalence, morbidity, and mortality of asthma are increasing in the United States. Puerto Ricans have the highest incidence, followed by non-Hispanic Blacks and Native Americans.

The incidence of asthma cases and asthma deaths is increasing nationwide, but the reason for the increase is unknown. Explanations include increased accuracy of medical diagnosis and increased chart documentation among physicians, as well as increased prevalence as average life expectancy increases.

Risk Factors

The environment, including air pollution and exposure to other environmental toxins (including pesticides),^{205,157} homes that are airtight, exposure to pets, and windowless offices may also be risk factors contributing to the significant rise in incidence. The *hygiene hypothesis* blames lack of exposure to stimulants or overexposure to cleaning agents.^{277,368}

Large families, early exposure to pets, early infections, and attending daycare may protect against allergic sensitization.⁴²⁶ Adults with asthma are 12 times more likely to develop COPD,¹⁸¹ and there are more than 4500 deaths from asthma each year in the United States.

Asthma can occur at any age, although it is more likely to occur for the first time before the age of 5 years. Antibiotic exposure during infancy appears to be a risk factor for developing childhood asthma. In childhood, it is three times more common and more severe in boys; however, after puberty, the incidence in the genders is equal, although monthly variations in asthma episodes seem to be correlated with estrogen levels for women.¹¹⁹

Children with lower birth weight (less than 5½ lb at birth) and prematurity (more than 3 weeks premature) are more susceptible to the effects of ozone (air pollution) compared with children who are born full term or full weight.³¹¹ It is estimated that asthma goes unrecognized as an adverse factor affecting performance in 1 in 10 adolescent athletes.

Asthma is found most often in urban, industrialized settings; in colder climates; and among the urban disadvantaged population (areas of poverty). Asthma is more prevalent and more severe among black children, but this may not be a result of race or low income per se but rather of demographic location because all children living in an urban setting are at increased risk for asthma.⁹ The number of African-American boys with asthma increased in 2001 to 2004, whereas the incidence remained stable for African-American girls and white children of both sexes.²⁷⁸

Overcrowded living conditions with repeated exposure to cigarette smoke, dust, cockroaches, and mold and where the use of a gas stove or oven is used for heat may be contributing factors.²⁵⁷ Alcoholic drinks, particularly wines, appear to be important triggers for asthmatic responses. Sensitivity to the sulfite additives and salicy-

lates present in wine seems likely to play an important role in these reactions.⁴²⁷

There is a relationship between obesity and asthma. Data from the Nurses Health Study II show that obesity increases women's risk of developing adult-onset asthma possibly as a result of estrogen stored in lipids. The higher the body mass index (BMI), the greater the risk of developing asthma.⁷² One study demonstrated reduced respiratory function in 32% of obese children and only 3% of children with a normal BMI.⁴²⁵

Inflammatory mediators are produced by adipose tissue and may represent an immunologic connection between asthma and obesity. Increased leptin secretion may enhance airway inflammation while reduced adiponectin (an antiinflammatory agent) secretion affects modulation of inflammation. Comorbidity of asthma and obesity may complicate the treatment of either condition and appropriate asthma treatment may help with exercise and weight loss. Prevention of obesity and/or weight loss should be encouraged for all persons with asthma.³⁴⁹

Etiologic Factors

Asthma occurs in families, which indicates that it is an inherited disorder. Asthma is influenced by two genetic tendencies: one associated with the capacity to develop allergies (atopy) and the other with the tendency to develop hyperresponsiveness of the airways independent of atopy. Eighty percent of individuals with asthma report allergic rhinitis. Studies have shown that stimulation of the nasal mucosa causes bronchi to react.³³⁶ Environmental factors interact with inherited factors to cause attacks of bronchospasm. Asthma can develop when predisposed persons are infected by viruses or exposed to allergens or pollutants.

Extrinsic asthma, also known as atopic or allergic asthma, is the result of an allergy to specific triggers; usually the offending allergens are foods or environmental antigens suspended in the air in the form of pollen, dust, molds, smoke, automobile exhaust, and animal dander. In this type of asthma, mast cells, sensitized by IgE antibodies, degranulate and release bronchoactive mediators after exposure to a specific antigen. More than one-half of the cases of asthma in children and young adults are of this type.

Intrinsic asthma, or nonallergic asthma, has no known allergic cause or trigger, has an adult onset (usually over 40 years of age), and is most often secondary to chronic or recurrent infections of the bronchi, sinuses, or tonsils and adenoids. This type of asthma may develop from a hypersensitivity to the bacteria, or more commonly, viruses causing the infection. Other factors precipitating intrinsic asthma include drugs (aspirin and P-adrenergic antagonists), environmental irritants (occupational chemicals and air pollution), cold dry air, exercise, and emotional stress.

Occupational asthma is defined as variable narrowing of airways, causally related to exposure in the working environment to specific airborne dusts, gases, acids, molds, dyes, vapors, or fumes. Many of these substances are very common and not ordinarily considered hazardous. Only a small proportion of exposed workers develop occupa-

tional asthma, but it has received considerable attention recently as the most frequent occupational lung disease worldwide.

New substances and processes involving new chemicals have increased dramatically in the last 2 decades, and there is little information about "safe" levels of exposure that protect all workers.²⁷ High-risk occupations for asthma include farmers, animal handlers, and agricultural workers; painters; plastics and rubber workers; cleaners and homemakers (especially if cooking is done with a gas stove); textile workers; metal workers; and bakers, millers, and other food processors. Exposure to biologic dusts and gases and fumes can cause a 30% to 50% increased risk of asthma.

Pathogenesis

The airways are the site of an inflammatory response consisting of cellular infiltration, epithelial disruption, mucosal edema, and mucous plugging (Fig. 15-11). The release of inflammatory mediators produces bronchial smooth muscle spasm; vascular congestion; increased vascular permeability; edema formation; production of thick, tenacious mucus; and impaired mucociliary function.

Several mediators also cause thickening of airway walls and increased contractile response of bronchial smooth muscles. These changes in the bronchial musculature, combined with the epithelial cell damage caused by eosinophil infiltration, result in the airway hyperresponsiveness characteristic of asthma.

Once the airway is in spasm and airways are swollen, mucus plugs the airway, trapping distal air. V/Q mismatch, hypoxemia, obstructed expiratory flow, and increased workload of breathing follow. Most attacks of asthmatic bronchospasm are short-lived, with freedom from symptoms between episodes, although airway inflammation is present, even in people who are asymptomatic.

Excessive airway narrowing occurs when the smooth muscle shortens (not necessarily to an abnormal degree). The relationship between the mechanical and contractile properties of smooth muscle and lung volume and how these interact to determine smooth muscle length are the subject of new research. The relative importance of smooth muscle area and mechanical properties, altered airway structure, and airway inflammation in asthma is not yet determined.²¹⁶

Although definitive causes of asthma have not been determined, there is much known about the immune system mechanisms that lead to allergic airway obstruction. T-helper cells secrete cytokine, which contribute to inflammation that is mediated by IgE. IgE is present on mast cells and other airway cells. After repeated contact with antigens, these cells break down and release toxins, particularly leukotrienes, which cause bronchospasm and hypersecretions.³⁰

Researchers are also investigating the possibility of underlying neurogenic mechanisms that may contribute to the pathogenesis and pathophysiology of asthma.^{88,361,336} Investigations include determination of the linkages among psychosocial factors and behavioral, neural, endocrine, and immune processes in the role of

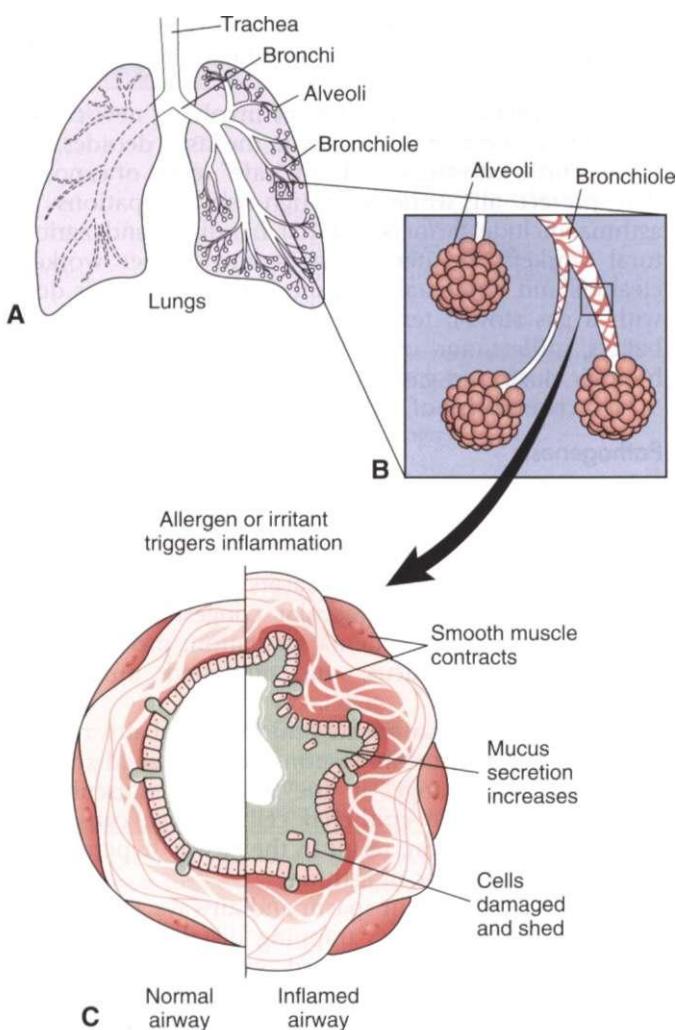


Figure 15-11

Bronchiole response in asthma. **A**, Air is distributed throughout the lungs via small airways called bronchioles. **B**, Healthy bronchioles accommodate a constant flow of air when open and relaxed. **C**, In asthma, exposure to an allergen or irritant triggers inflammation, causing constriction of the smooth muscle surrounding the bronchus (bronchospasm). The airway tissue swells; this edema of the mucous membrane further narrows airways with production of excess mucus also interfering with breathing.

asthma. It has been shown that health-related behaviors, demographic factors, and psychosocial factors influence susceptibility to and severity of exacerbation of asthma.^{240,390}

Clinical Manifestations

Clinical signs and symptoms of asthma differ in presentation (Box 15-6), degree (Table 15-8), and frequency among clients, and although current symptoms are the most important concern of affected people, they reflect the current level of asthma control more than underlying disease severity.³²⁷

During full remission, clients are asymptomatic and pulmonary function tests are normal. Over time, repeated attacks cause airway remodeling, chronic air trapping,

Box 15-6

CLINICAL MANIFESTATIONS OF BRONCHIAL ASTHMA

Cough

Hacking, paroxysmal, exhausting, irritative, involuntary, nonproductive
Becomes rattling and productive of frothy, clear, gelatinous sputum
Main or only symptom
Tickle in the back of the throat accompanied by a cough

Respiratory-Related Signs

Shortness of breath (SOB); may occur at rest
Prolonged expiratory phase
Audible wheeze on inspiration and expiration or on expiration only; never on inspiration only
Often appears pale
May have a malar flush and red ears
Lips deep dark red color
May progress to cyanosis of nail beds, around mouth and lips
Restlessness
Apprehension
Anxious facial expression
Itching around nose, eyes, throat, chin, scalp
Sweating may be prominent as attack progresses
May sit upright with shoulders in a hunched-over position, hands on the bed or chair, and arms braced (older children)
Speaks with short, panting broken phrases

Chest

Coarse, loud breath sounds (may become quiet or silent if severe)
Prolonged expiration
Generalized inspiratory and expiratory wheezing; increasingly high-pitched
Loss of breath sounds with severe cases

With Repeated Episodes

Barrel chest
Elevated shoulders
Use of accessory muscles of respiration
Skin retraction (clavicles, ribs, sternum)
Facial appearance: flattened malar bones, circles beneath the eyes, narrow nose, prominent upper teeth, nostrils flaring

Modified from Wong DL: *Whaley and Wong's essentials of pediatric nursing*, ed 5, St Louis, 1997, Mosby-Year Book.

proliferation of submucosal glands, and hypertrophied smooth muscle. This may progress to irreversible changes and COPD.

At the beginning of an attack, there is a sensation of chest constriction, inspiratory and expiratory wheezing, nonproductive coughing, prolonged expiration, tachycardia, and tachypnea. Secondary bronchospasm is marked by recurrent attacks of dyspnea, with wheezing caused by the spasmodic constriction of the bronchi. Other symptoms may include fatigue, a tickle in the back of the throat accompanied by a cough in an attempt to clear the airways, and nostril flaring (advanced).

The person usually assumes a classic sitting or squatting position to reduce venous return, leaning forward so

Table 15-8 Stages of Asthma

Stage	Symptoms
Mild	Symptoms reverse with cessation of activity; daytime symptoms ≤2 times/wk; nighttime symptoms ≤2 times/mo; inhaled medication as needed (not usually daily)
Moderate	Audible wheezing Use of accessory muscles of respiration Leaning forward to catch breath Daily (but not continual) daytime symptoms requiring short-acting inhalant and long-term treatment Episodes ≥2 times/wk; nighttime symptoms ≥4 times/mo
Severe	Blue lips and fingernails Tachypnea (30-40 breaths/min) despite cessation of activity Cyanosis-induced seizures Skin and rib retraction Activity limited; frequent daytime and nighttime episodes, sometimes continual

as to use all the accessory muscles of respiration. The skin is usually pale and moist with perspiration, but in a severe attack there may be cyanosis of the lips and nail beds. In the early stages of the attack coughing may be dry, but as the attack progresses, the cough becomes more productive of a thick, tenacious mucoid sputum. The nocturnal worsening of asthma is a common feature of this disease and may affect daytime alertness, even in children.¹¹⁶ Phinitis, chronic cough, snoring, and apnea may be responsible for sleep disturbance.

An acute attack that cannot be altered with routine care is called *status asthmaticus*. This is a medical emergency requiring more vigorous pharmacologic and support measures. Despite appropriate treatment, this condition can be fatal. With severe bronchospasm, the workload of breathing increases 5 to 10 times, which can lead to acute cor pulmonale.

When air is trapped, a severe paradoxical pulse develops as venous return is obstructed; blood pressure drops over 10 mm Hg during inspiration. Pneumothorax occasionally develops. If *status asthmaticus* continues, hypoxemia worsens and acidosis begins. If the condition is untreated or not reversed, respiratory or cardiac arrest will occur.

MEDICAL MANAGEMENT

PREVENTION. Heavier emphasis on teaching self-management and especially prevention for anyone with asthma is recommended by the American Academy of Allergy, Asthma, and Immunology. An excellent daily asthma management plan is available.¹¹ *Healthy People 2010* has identified 11 objectives specifically related to this condition, including increasing the proportion of people with asthma who receive formal education as part of their management program (see *Healthy People 2010*: <http://health.gov/healthypeople/>).

DIAGNOSIS. Pulse oximetry, pulmonary function studies, bronchial challenge test with methacholine, skin prick tests, ABG analysis, serum IgE and blood eosinophil counts, induced sputum cell counts, exhaled nitric oxide (marker for eosinophilic inflammation),⁴¹⁹ questionnaires, and chest films may be used in assessing for both the presence and the severity of asthma (see Chapter 40 for a description of and reference values for these tests). The methacholine challenge test is a valuable diagnostic measure. There are strong correlations with this test and some symptoms.⁴⁶² Inexpensive but reliable spirometer testing can be used to obtain evidence of the bronchial hyperreactivity associated with asthma.

Diagnosis may be delayed in older clients who have other illnesses that cause similar symptoms or who attribute their breathlessness to the effects of aging and respond to the onset of asthma by limiting their activities to avoid eliciting symptoms. The diagnosis of occupational asthma is usually based on history of a temporal association between exposure and the onset of symptoms and objective evidence that these symptoms are related to airflow limitation. Sputum induction and analysis may be helpful in confirming occupational asthma.¹⁵⁹

TREATMENT. Identifying specific allergens for each individual and avoidance of asthma triggers, combined with the use of two classes of medications (bronchodilators and antiinflammatory agents; see Table 15-6), has been recommended in the management of asthma. In the past, asthma was thought to be caused by spasms of the muscles surrounding the airways between the trachea and lungs and therefore treated first with bronchodilator drugs to widen the constricted airways and ease symptoms. It is now clear that asthma attacks are actually episodic flareups of chronic inflammation in the lining of the airways necessitating the use of inhaled antiinflammatories to suppress the underlying inflammation and allow the airways to heal.

Most people require bronchodilator therapy to control symptoms by activating β-agonist receptors on smooth muscle cells in the respiratory tract, thereby relaxing the bronchial muscle and opening the airways; those with mild symptoms may use metered-dose inhaler (MDI) devices to administer sympathomimetic bronchodilators on an as-needed basis.

People who experience moderate-to-severe asthma may require daily administration of antiinflammatory agents, such as corticosteroids, to prevent asthma attacks. Corticosteroids dampen the entire immune system response. Antiinflammatory drugs have a preventive action by interrupting the development of bronchial inflammation. They may also modify or terminate ongoing inflammatory reactions in the airways.

It is important that people with asthma know the difference between medications that must be taken daily to prevent asthma symptoms and medications that relieve symptoms once they begin. Low-dose corticosteroid inhalants are recommended to reduce the risk of side effects (e.g., psychiatric problems, reduced growth in children, ocular effects, death, osteoporosis, or alopecia and hirsutism) from prolonged use.^{117,407} Leukotriene-

receptor antagonists inhibit inflammation and have been shown to be safe and effective in adults with asthma and allergic rhinitis⁴³⁴ (see further discussion in Chapter 6).

Oxygen metabolites (free radicals) may play a direct or indirect role in the modulation of airway inflammation. Excessive superoxide and hydroxyl radical production accompanied by significantly lower free radical scavengers in asthma (the latter even during rest) endorses the correlation between disease severity and oxygen radical production in people with asthma.³⁸³

Low dietary intake and blood levels of vitamins C and E, selenium, and flavonoids are seen in people with asthma.^{302,337,384} Although some researchers suggest that antioxidant nutrients (especially obtained from food sources such as fruits and vegetables) appear helpful in asthma treatment,^{288,300,317,337} others report that people with asthma may have a diminished capacity to restore the antioxidant defenses, making the use of supplemental antioxidants questionable in this population.³⁴¹

Many complementary treatments have been used to ameliorate asthma symptoms though there has been minimal research to validate most of these claims. There is minimal but growing evidence to support the use of acupuncture.²⁶⁶ Spinal manipulation and other manual therapy have been deemed to be ineffective in the treatment of asthma.^{128,203} In one study of 65 people, yoga was found to have no effect on asthma.³⁷⁰ Complementary treatments are still being studied for benefits and adverse affects.^{42b}

Genetic treatment is under investigation. For example, gene transfer into the airway cells to block mediator proteins (signal transducer activator of transcription [STAT]) from setting off an immune response or too strong an immune response in the asthma pathway is the subject of several studies.^{129,372} A recombinant monoclonal antibody (Omalizumab) was approved by the FDA in 2003. This antibody prevents IgE from binding to mast cells and other effector cells, preventing inflammation.⁴³⁸

PROGNOSIS. The outlook for clients with *bronchial asthma* is excellent despite the recent increase in the death rate. Childhood asthma may disappear, but only about one-quarter of the children with asthma become symptom-free when their airways reach adult size. Factors that predict adult asthma include gender (males are more likely to outgrow asthma), smoking, allergy to dust mites, degree of airway hyperresponsiveness, and early age of onset.^{379,418} Adults with asthma are 12 times more likely to progress to COPD, but studies indicate that the majority of people with asthma do not experience a decline in pulmonary mechanics or appear to be at risk for reduced life expectancy.^{181,1290}

Attention to general health measures and use of pharmacologic agents permit control of symptoms in nearly all cases. The risk of lung cancer is two times greater in people with asthma compared with those who do not have a history of asthma.⁶⁶

Status asthmaticus can result in respiratory or cardiac arrest and possible death (see previous discussion). If ventilation becomes necessary, prognosis for recovery is poor.^{181,1290}

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-7

Asthma

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Many people with asthma do not even know they have the disease. Some think they simply have chronic bronchitis, colds, or allergies. Anyone who reports coughing or a feeling of tightness in the chest when others smoke nearby and especially anyone who gasps for breath after exercise should be referred to a physician for evaluation of these symptoms.

Exercise-Induced Asthma

Exercise-induced bronchospasm (EIB), or exercise-induced asthma (EIA), does not represent a unique syndrome but rather an example of the airway hyperactivity common to all persons with asthma. EIA is an acute, reversible, usually self-terminating airway obstruction that develops 5 to 15 minutes after strenuous exercise when the person no longer breathes through the nose, warming and humidifying the air, but opens the mouth. Breathing cold, dry air through the mouth degranulates mast cells that release bronchoconstrictive mediators inducing EIB. EIB/EIA lasts 15 to 60 minutes after the onset.

Because physical therapists prescribe and observe exercise, they may be the first to recognize symptoms of undiagnosed asthma. Coughing is the most common symptom of EIA, but other symptoms include chest tightness, wheezing, and SOB. The affected (but undiagnosed) individual may comment, "I am more out of shape than I thought." This should be a red flag for the therapist to consider the possibility of asthma and need for medical diagnosis and intervention.

If an asthma attack should occur during therapy, first assess the severity of the attack. Place the person in the high Fowler's position and encourage diaphragmatic and pursed-lip breathing. If the client has an inhaler available, provide whatever assistance is necessary for that person to self-administer the medication. Help the person relax while assessing the person's response to the medication.

Usually the episode subsides spontaneously in 30 to 60 minutes. The severity of an attack increases as the exercise becomes increasingly strenuous. Warmup exercise can ameliorate the inflammatory process by affecting platelet-eosinophil/neutrophil reactions.⁴³⁹ The problem is rare in activities that require only short bursts of energy (e.g., baseball, sprints, gymnastics, or skiing) compared with those that involve endurance

Table 15-9 Exercise Guidelines for Children With Asthma

Recommendation	Benefit
General exercise, school-based physical education.	Maintains motor control, flexibility, strength, and cardiovascular fitness and prevents or reverses side effects of medication (e.g., corticosteroids).
Low-impact exercise (aerobics, weight training, stationary bike). Warmup before aerobic activity.	Raises threshold for strenuous exercise before mouth breathing and EIB occur.
Exercise in a trigger-free environment (i.e., avoid cold, pollution, or increased pollen outdoors; exercise indoors; avoid tobacco smoke; swimming program is ideal).	Permits exercise without increased bronchospasm.
Take prescribed medication properly before exercise or activity producing bronchospasm.	Helps control airway reactivity; gradually desensitizes mast cells, reducing release of bronchoconstrictive mediators.
Monitor FEV ₁ /FVC ratio before, during, and after physical activity.* Decrease of 10% requires slowing activity. Drop of 15%-20% from initial measurement requires cessation of exercise.	Prevents bronchospasm; controls symptoms.

EIB, Exercise-induced bronchospasm; FEV₁, ratio of forced expiratory volume in 1 sec to forced vital capacity.

*Peak flowmeters can be used to obtain this information. Determine the child's normal range of lung function by having the child blow in the meter in the morning and evening for 1 week. The average level measured varies from person to person and is influenced by gender and height. Testing should establish a peak flow protocol against which lung function can be compared to determine if deterioration has occurred.

exercise (e.g., soccer, basketball, distance running, or biking).

Swimming, even long-distance swimming, is well tolerated by people with EIA, partly because they are breathing air fully saturated with moisture, but the type of breathing required may also play a role. Exhalation under water, which is essentially pursed-lip breathing, is beneficial because it prolongs each expiration and increases the end-expiratory pressure within the respiratory tree.

EFFECT OF EXERCISE

There are many barriers to exercise for people with asthma, including lack of motivation, time constraints, weather conditions, and belief that exercise is not good for this condition.²⁷³ To prevent secondary complication of a sedentary lifestyle and because obesity can contribute to the inflammatory process associated with asthma, exercise and education about exercise should be part of any treatment program.

There is strong evidence to support physical training for cardiovascular training in this population.³⁵¹ There is inadequate evidence for a positive effect of breathing exercises and inspiratory muscle training in individuals who have asthma.^{202,352}

Exercise and Medication

Bronchospasm can occur during exercise (especially in EIA) if the person with asthma has a low blood oxygen level before exercise. For this reason, it is helpful to take bronchodilators by MDI 20 to 30 minutes before exercise, performing mild stretching and warmup exercises during that time period to avoid bronchospasm with higher workload exercise. Increased exercise should be accompanied by good bronchodilator coverage to promote bronchodilation and improve alveolar venti-

lation and oxygenation. Exercise guidelines for adults with asthma can be modified from recommendations for children with asthma (Table 15-9).

Many clients have found that using their inhalers in this way before exercise permits them to exercise without onset of symptoms. Proper administration of an MDI is essential (see Box 15-5). The first dose induces dilation of the larger, central bronchial tubes, relaxing smooth muscles in the airways; the second dose dilates the bronchioles (smaller airways).

Metabolism of certain drugs administered can be altered by exercise, tobacco, marijuana, or phenobarbital (all of which increase drug metabolism). Cimetidine (Tagamet), erythromycin, or the presence of a viral infection may decrease drug metabolism.

If a client develops signs of asthma or any bronchial reactivity during exercise, the physician must be informed. Medication dosage can then be altered to maintain optimal physical performance. Excessive use of inhaled P-adrenergic agents (using three or more full canisters monthly) requires physician referral for further evaluation.

Common manifestations of drug-induced (theophylline) toxicity include nausea, vomiting, tremors, anxiety, tachycardia and arrhythmias, and hypotension. The use of nonsteroidal antiinflammatory drugs (NSAIDs), including aspirin, in older people with asthma should be avoided if possible because the drug interactions can cause increased bronchospasm in susceptible individuals.

Some athletes do not achieve the control needed for the performance demands of competition. The effectiveness of short-acting medications and medications in general for asthma varies widely among people with asthma while exercising. The preventive benefits of

Continued.

each medication dose may wane after taking a new drug for several weeks. Any athlete with asthma who cannot perform at the levels desired or expected because of asthma symptoms should be advised to review medications and medication use with the physician.

Medication and Bone Density

Long-term use of inhaled corticosteroids in the management of moderate-to-severe asthma is associated with decreased bone mineral density (BMD) and associated increased risk of fractures and a high occurrence of asymptomatic vertebral fractures, particularly in high-risk postmenopausal women (i.e., those not receiving hormone replacement therapy).^{15,364,451} African-American children may be afforded some protection from osteoporosis as compared to Caucasian children using high-dose inhaled corticosteroids.¹⁷⁶

All people receiving glucocorticoid therapy (e.g., prednisolone) at doses of 7.5 mg/day or more for 6 months or longer should discuss with their physician the use of low-dose inhaled corticosteroids, assessment of BMD, and preventive therapies (e.g., bisphosphonates, calcium supplementation, or vitamin D).³²¹ The physical therapist can be very instrumental in providing education for the prevention and intervention for the treatment of osteopenia and osteoporosis. See the section on Osteoporosis in Chapter 24.

Monitoring Vital Signs

Monitoring vital signs can alert the therapist to important changes in bronchopulmonary function. Developing or increasing tachypnea may indicate worsening asthma or drug toxicity. Other signs of toxicity, such as diarrhea, headache, and vomiting, may be misinterpreted as influenza. Hypertensive blood pressure readings may indicate asthma-related hypoxemia.

Auscultate the lungs frequently, noting breath sounds including the degree of wheezing and the quality of air movement. In this way, any change in respiratory status will be more readily perceived. If the client does not have a productive cough in the presence of rhonchi (dry rattling in the bronchial tube), teach effective coughing techniques.

Status Asthmaticus

Therapy can augment the medical management of the client with status asthmaticus. In coordination with the individual's medications, the therapist helps to remove secretions; promotes relaxed, more efficient breathing; enhances V/Q matching; reduces hypoxemia; and teaches the client to coordinate relaxed breathing with general body movement.

Caution needs to be observed to avoid stimuli that bring on bronchospasm and deterioration (e.g., aggressive percussion, forced expiration maneuvers, aggressive bag ventilation, or manual hyperinflation with an intubated individual). Certain body positions may have to be avoided because of client intolerance or exacerbation of symptoms in those positions.¹¹¹

Immediate medical care is recommended for anyone with asthma who is struggling to breathe with no

improvement in 15 to 20 minutes after initial treatment with medications or who is hunched over and unable to straighten up or resume activity after medication dosage. The presence of blue or gray lips or nail beds is another indication of the need for immediate medical attention.

Bronchiectasis

Definition

Bronchiectasis is a progressive form of obstructive lung disease characterized by irreversible destruction and dilation of airways generally associated with chronic bacterial infections. Clinically, it is considered an extreme form of bronchitis and no longer considered part of COPD. Abnormal and permanent dilation of the bronchi and bronchioles develops when the supporting structures (bronchial walls) are weakened by chronic inflammatory changes associated with secondary infection.

Incidence and Etiologic and Risk Factors

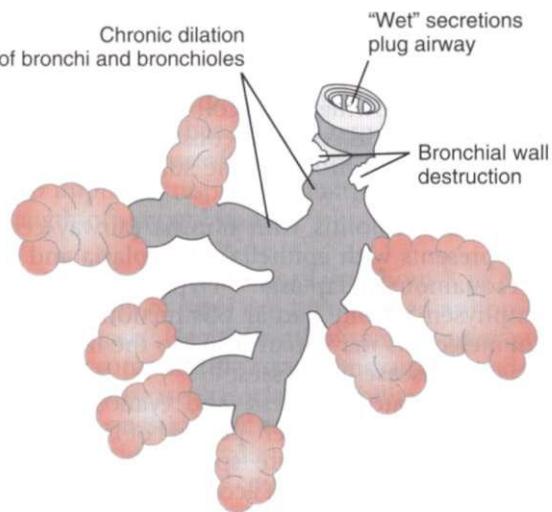
The incidence of bronchiectasis is low in the United States because of improved control of bronchopulmonary infections. However, any condition producing a narrowing of the lumen of the bronchioles may create bronchiectasis, including TB, adenoviral infections, and pneumonia. Bronchiectasis also develops in people with immunodeficiencies involving humoral immunity, recurrent aspiration, and abnormal mucociliary clearance (immotile cilia syndromes).

CF causes about one-half of all cases of bronchiectasis. Sinusitis, dextrocardia (heart located on right side of chest), Kartagener's syndrome (alterations in ciliary activity), defective development of bronchial cartilage (Williams-Campbell syndrome), and endobronchial tumor predispose a person to bronchiectasis.

Pathogenesis

Although bronchiectasis has been viewed as a progressive disease of destruction and dilation of the medium and large airways, there is now evidence of the importance of the small airways in the pathogenesis of this condition. Chronic inflammation of the bronchial wall by mononuclear cells is common to all types of bronchiectasis. Abnormal bronchial dilation characteristic of bronchiectasis is accompanied by accumulation of wet secretions that plug the airway and cause bronchospasm, producing even more mucus. A vicious cycle of bacteria-provoked inflammatory lung damage occurs with irreversible destruction or fragmentation of the bronchial wall and resultant fibrosis further obstructing and obliterating the bronchial lumen (Fig. 15-12).

In response to these changes, large anastomoses develop between the bronchial and pulmonary blood vessels to increase the blood flow through the bronchial circulation. V/Q mismatch causes hypoxia and hypercapnia. Damage to these anastomoses is responsible for the hemoptysis present in persons with bronchiectasis.

**Figure 15-12**

Airway pathology in bronchiectasis.

Clinical Manifestations

The most immediate symptom of bronchiectasis is persistent coughing, with large amounts of purulent sputum production (worse in the morning). Rhinosinusitis, dyspnea, and fatigue are also strongly related to bronchiectasis. For those without other significant disease, most report chronic childhood respiratory problems.²⁴²

Weight loss, anemia, and other systemic manifestations, such as low-grade fever, hemoptysis, and weakness, are also common. Clubbing may occur, and the breath and sputum may become foul-smelling with advanced disease. Heart failure may occur due to the vascular fibrosis. There is a known correlation between bronchiectasis and rheumatoid arthritis, but the exact mechanism remains unknown.

MEDICAL MANAGEMENT

DIAGNOSIS. Imaging studies (e.g., high-resolution CT scan) have become increasingly accurate in depicting the features of early bronchiectasis. Ultrafast and multislice CT scans have increased the value of CT as a diagnostic tool by reducing the need for sedation or anesthesia. Other diagnostic tests include radiographic studies, laboratory tests including sweat chloride test for CF, electron microscopy of bronchial biopsy for immotile cilia, and sputum culture analyses.

TREATMENT. The goals of treatment are removal of secretions and prevention of infection. The principal treatment is airway clearance techniques, bronchodilators, and antibiotics selected on the basis of sputum smears and cultures. A recent study demonstrated that use of inhaled corticosteroids improved quality of life for people with steady-state bronchiectasis.²⁸² Hydration is important, and oxygen may be administered. Surgical resection is reserved for the few clients with localized bronchiectasis and adequate pulmonary function who fail to respond to conservative management or for the person with massive hemoptysis. Long-term care is the same as for any person with COPD.

PROGNOSIS. The morbidity and mortality associated with bronchiectasis have declined markedly in industrialized nations, but prevalence remains high in Pacific and Asian countries. The overall prognosis is often poor, and although bronchiectasis is usually localized to a lung lobe or segment, persistent, nonresolving infection may cause the disorder to spread to other parts of the same lung. Complications of bronchiectasis include recurrent pneumonia, lung abscesses, metastatic infections in other organs (e.g., brain abscess), and cardiac and respiratory failure. Good pulmonary hygiene and avoidance of infectious complications in the involved areas may reverse some cases of bronchiectasis.

SPECIAL IMPLICATIONS FOR THE THERAPIST

15-8

Bronchiectasis

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

The effects of bronchopulmonary airway clearance techniques (formerly referred to as *hygiene physical therapy, chest therapy, chest physical therapy*) to improve pulmonary function in bronchiectasis remain inconclusive because of insufficient research.^{228,229} The beneficial effects of airway clearance techniques to mobilize secretions and improve pulmonary clearance (e.g., sputum production or radioaerosol clearance) in the treatment of bronchiectasis have been documented in one small study.²²⁹

In many settings, postural drainage and percussion for the person with bronchiectasis is administered routinely on the basis of diagnosis rather than specific clinical criteria. Further research to clarify outcomes of airway clearance techniques is necessary and may provide the therapist with clinical goals other than secretion mobilization.

Inspiratory muscle training appears to have an effect on exercise endurance, but insufficient evidence exists to support other types of physical training.⁵⁶ However, a recent randomized control study demonstrated improvement in exercise tolerance with pulmonary rehabilitation.⁵⁵

The selection of techniques to include in an airway clearance regimen varies among institutions as well as among practitioners and may include positioning, postural drainage, and chest percussion of involved lobes performed several times per day. Family members can be instructed in how to provide this care at home.

Directed coughing and breathing exercises to promote good ventilation and removal of secretions should follow positional or percussive therapy. The

Continued.

best times to do this are early morning and several hours after eating the final meal; performing these techniques just before bedtime may result in increased coughing and prevent the person from sleeping. An excellent review of the use of airway clearance techniques in the acute care setting is available.¹⁴⁰

Bronchiolitis

Definition and Overview

Bronchiolitis refers to several morphologically distinct pathologic conditions that involve the small airways. Acute bronchiolitis is a commonly occurring, diffuse, and often severe inflammation of the lower airways (bronchioles) in children under 2 years caused by a viral infection. Acute adult onset is related to asthma, aspiration, or bronchiectasis.

Bronchiolitis was once classified as a type of chronic interstitial pneumonia and referred to as *small airways disease*; progress in pathology has provided more specific etiology-directed diagnoses that reflect the individual reaction patterns observed. Constrictive bronchiolitis, diffuse panbronchiolitis, and airway-centered fibrosis are also forms of bronchiolitis.

Bronchiolitis obliterans in the adult is now considered acute or chronic with identification of these special forms (e.g., obliterative, eosinophilic bronchiolitis in asthma, necrotizing bronchiolitis in viral infection, or toxic fume bronchiolitis after exposure to noxious gases and the development of chemical pneumonitis).^{346,435} Bronchiolitis obliterans is the most important clinical complication in heart-lung and lung transplant recipients and may represent a form of allograft rejection; it is a rare complication of allogeneic (human-to-human) bone marrow transplantation. Circulating fibroblast precursors from bone marrow may be implicated in transplant recipients with bronchiolitis.⁶³

Bronchiolitis obliterans may occur in association with rheumatoid arthritis, polymyositis, and dermatomyositis. Penicillamine therapy has been implicated as a possible cause of bronchiolitis obliterans in clients with rheumatoid arthritis.

Incidence and Etiologic Factors

Bronchiolitis obliterans in adults usually occurs with chronic bronchitis; bronchiolitis in children is associated with pulmonary infections, such as respiratory syncytial virus (RSV), parainfluenza viruses, adenoviruses, or pertussis (whooping cough), or associated with measles. Primarily present in winter and spring, it is easily spread by hand-to-nose or nose-to-eye transmission. Exudative bronchiolitis, which is inflammation of the bronchioles with exudation of grey tenacious sputum, is often associated with asthma.

Pathogenesis and Clinical Manifestations

Variable degrees of obstruction occur in response to infection as the bronchiolar mucosa swells and the lumina fill with mucus and exudate. Depending on the type, these changes occur as the walls of the bronchi and bronchioles

are infiltrated with inflammatory cells, increased goblet cells, and fibroblasts.

Bronchiolitis obliterans is usually confined to one lobe and its airways, showing dense collagen but no proliferation of fibroblasts. In contrast, constrictive bronchiolitis is characterized by fibrosis of the submucosa and peribronchial tissues.

Chronic bronchiolitis with fibrosis (airway centered fibrosis) presents with epithelial hyperplasia and goblet cell and squamous metaplasia.⁴³⁵ Hyperinflation, obstructive emphysema from partial obstruction, and patchy areas of atelectasis may occur distal to the inflammatory lesion as the disease progresses.

Cough, respiratory distress, and cyanosis occur initially, followed by a brief period of improvement. Dyspnea, paroxysmal cough, sputum production, and wheezing with marked use of accessory muscles follow as the disease progresses. Apnea may be the first indicator of RSV infection in very young infants. Severe disease may be followed by a rise in Paco_2 (hypercapnia), leading to respiratory acidosis and hypoxemia.

MEDICAL MANAGEMENT

PREVENTION. Currently there is no vaccine for RSV. The outcome of infection depends on host and viral genetics. A better understanding of RSV molecular biology and pathogenesis will help facilitate an effective vaccine and molecular targets for new drug treatment. Understanding RSV disease mechanisms in order to develop a vaccine is difficult because there is a wide range of RSV disease phenotypes in humans and disparities in RSV disease phenotypes among the animal models used in research.³⁰⁸

Frequent handwashing and not sharing items, such as cups, glasses, and utensils, with persons who have RSV illness can decrease the spread of virus to others. Excluding children with colds or other respiratory illnesses (without fever) who are well enough to attend childcare or school settings may not decrease the transmission of RSV, since it is often spread in the early stages of illness. In a hospital setting, RSV transmission can and should be prevented by strict attention to contact precautions such as handwashing and wearing gowns and gloves.

DIAGNOSIS AND TREATMENT. Diagnosis is made on the basis of clinical findings, age, the season, and the epidemiology of the community. On chest radiographs, this condition is difficult to differentiate from bacterial pneumonia. RSV can be positively identified using an enzyme-linked immunosorbent assay (ELISA) from direct aspiration of nasal secretions. Early diagnosis in transplant recipients may be facilitated by using CT.¹¹²

There is no specific treatment for bronchiolitis, and medical therapy is controversial. Treatment modalities may include steroids, humidified air, hydration, and airway clearance techniques such as postural drainage, coughing, and deep-breathing exercises. Antibiotics may be used initially when a bacterial cause of illness has not been ruled out or for secondary infections. Mist therapy combined with oxygen by hood or tent to alleviate dyspnea and hypoxia may be used with children.

Corticosteroids should be used with caution because there is a relationship between levels of Cortisol and severity of the disease.³⁴³ Recently, heliox therapy (combination of oxygen and helium) has been shown to have a positive effect on wheezing and respiratory distress.⁷³ In another study, the use of hypertonic saline with epinephrine also improved respiratory status and decreased hospital length of stay of infants.⁴¹⁴

PROGNOSIS. The acute disease lasts about 3 to 10 days, and the majority of cases can be managed at home with a good prognosis. Hospitalization may be necessary for anyone with complicating conditions such as underlying lung or heart disease, associated debilitated states, poor hydration, or questionable care at home. Some children deteriorate rapidly and die within weeks; others may follow a more long-term course. Onset before the age of 1 may be related to allergies or asthma, but there appear to be adverse long-term pulmonary consequences.⁴¹² In the adult, the acute form usually has a good prognosis, but the prognosis for chronic bronchiolitis obliterans is poor.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-9

Bronchiolitis

PREFERRED PRACTICE PATTERNS

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

RSV is the most common cause of pediatric acute bronchiolitis and pneumonia (see the section on RSV in Chapter 8). For this reason, any staff member with evidence of URI serves as a potential reservoir of RSV and should be excluded from direct contact with high-risk infants. All persons who come within 3 feet of an RSV client must wear a gown and mask with an eye shield and keep hands away from the face, especially the eyes, nose, and mouth. Hands must be washed before and after caring for any client and after handling potentially contaminated client care equipment. Standard Precautions (see Appendix A) must be strictly carried out.

Because RSV is readily transmitted by close contact with personnel, families, and other children both by direct contact (especially lifting or holding) and through contact with objects handled by the child, precautions against cross-infection are important. The primary routes of inoculation for the organisms are large-droplet inhalation through the nose and eyes. When contact is made with mucous discharge or drainage from the eye, nose, or mouth, the therapist is reminded to wear an exterior hospital gown and to discard the gown (or change clothing) when leaving.

Pregnant female personnel or visitors should be advised of the risk of potential physical defects in the

developing embryo from contact with RSV. Immunoprophylaxis to prevent RSV in high-risk infants is effective, and prevention of RSV may be possible in the future with active maternal immunization during pregnancy providing passive immunity of infants.^{161,431}

Sleep-Disordered Breathing

Definition

Sleep-disordered breathing comprises a collection of syndromes characterized by breathing abnormalities during sleep that result in intermittently disrupted gas exchange and in sleep interruption. Sleep-disordered breathing includes CSR, hypoventilation syndromes with and without chronic lung disease, heavy snoring with daytime sleepiness (upper airway resistance syndrome), and sleep apnea. The most common and only one discussed here, sleep apnea syndrome, is defined as significant daytime symptoms (e.g., sleepiness) in conjunction with evidence of sleep-related upper airway obstruction and sleep disturbance.⁴¹

There are three types of sleep apnea: central, obstructive, and mixed. *Central sleep apnea* is caused by altered chemosensitivity and cerebral respiratory control. In this type of apnea, the brain fails to send the appropriate signals to the respiratory muscles to initiate breathing and there is no diaphragmatic movement and no airflow. This is seen in infants less than 40 weeks' conceptual age and in people with neurologic disorders (e.g., tumors, brain infarcts, diffuse encephalopathies).

The most dramatic presentation is the person with repetitive apneas during sleep, accompanied by extreme daytime sleepiness. *Obstructive sleep apnea* (OSA), the most commonly diagnosed form of sleep apnea, is characterized by respiratory effort without airflow because of upper airway obstruction. *Mixed sleep apnea* is a central apnea that is immediately followed by an obstructive event.

Incidence

Depending on geographic area, prevalence of OSA ranges from 2% to 5.8% in men and 10% to 37% in women.³⁴⁹ Gender differences in upper airway collapse have not been observed, although there appears to be a relationship to testosterone levels.^{366,465,466} Forty percent of obese people have OSA, and 70% of people with sleep apnea are obese. OSA in children without neurologic impairment is most commonly caused by adenotonsillar hypertrophy, although obesity is positively correlated with this disorder in children also.¹⁸ Children with Down syndrome are particularly vulnerable to OSA and should be tested at age 3 to 4 years.³⁸⁷

Etiologic and Risk Factors

OSA is due to partial or complete pharyngeal collapse during sleep, leading to either reduction (hypopnea) or cessation (apnea) of breathing. The main cause in adults is upper body obesity, especially a large neck circumfer-

ence. A neck circumference greater than 16 inches for a woman or greater than 17 inches in a man correlates with an increased risk for this disorder.⁴³²

People with anatomically narrowed upper airways, such as occur in micrognathia; macroglossia (large tongue); and adenoid, uvula, elongated soft palate, or tonsillar hypertrophy, are predisposed to the development of OSA. Fat deposits or swelling in any or all of these tissues causes further obstruction. Other risk factors include increasing age; genetic factors (sleep-disordered breathing clusters in families); neurologic disorders; smoking; and cardiopulmonary dysfunctions such as hypertension, moderate-to-severe heart failure (including cor pulmonale), calcification of carotid arteries, chronic bronchitis, or cardiac dysrhythmia. Alcohol or sedatives before sleeping may precipitate or worsen the condition.

Pathogenesis

There are several hypotheses as to the pathogenesis of sleep apnea syndrome. Collapse or obstruction of the airway may occur with the inhibition of muscle tone that characterizes rapid eye movement (REM) sleep. When loss of normal pharyngeal muscle tone allows the larynx to collapse passively during inspiration, upper airway obstruction occurs and prevents effective ventilation.

By definition, apnea is a complete cessation of ventilation and therefore is precipitated by complete pharyngeal collapse, whereas hypopnea results from partial pharyngeal closure and is manifested by a substantial reduction in, but not a cessation of, breathing. Both conditions can lead to substantial hypoxia and hypercapnia with arousal from sleep required to reestablish airway patency and a resumption of ventilation. This cycle of recurrent pharyngeal collapse with subsequent arousal from sleep leads to the primary symptoms of daytime somnolence.¹³⁵ Enlarged tonsils or adenoids are the primary cause of sleep apnea in children.³⁵³

Clinical Manifestations

The frequent interruptions of deep, restorative sleep often lead to daytime (including morning) sluggishness and headaches, daytime fatigue, excessive daytime sleepiness, cognitive impairment, recent weight gain, and sexual impotence. Bed partners usually report loud cyclic snoring with periods of silence (breath cessation), restlessness, frequent episodes of waking up gasping, and often thrashing movements of the extremities during sleep.

Neurocognitive effects may include personality changes; irritability, hyperactivity, and depression; judgment impairment or poor school performance; domestic, work-related, or automobile accidents; memory loss; and difficulty concentrating.

Neurocognitive effects of apnea may also cause a mood disorder leading to an erroneous diagnosis of dysthymia; treatment with standard antidepressant medications may exacerbate the condition.²³³ Anyone with acquired or congenital neurologic disorders, such as tetraplegia, stroke, and Down syndrome, is susceptible to sleep disorders.^{387,401}

Fragmented sleep with its repetitive cycles of snoring, airway collapse, and arousal may cause hypertension in

some people. Sleep-disordered breathing may be a risk factor for cardiovascular involvement, including angina pectoris, acute myocardial infarction, cardiac arrhythmias, and ischemic stroke.^{64,65,338} The exact mechanisms for this are yet unknown, but there is an association with inflammation and prothrombic factors, as well as CNS effects.²³⁷ Risk for stroke is independent of other risk factors, including obesity and hypertension.⁴⁵⁷

MEDICAL MANAGEMENT

DIAGNOSIS. Diagnosis may be made using sleep monitoring devices, radiologic imaging, laboratory assays, questionnaires, and clinical signs and symptoms, but the most reliable test to confirm the diagnosis is overnight polysomnography (i.e., monitoring the subject during sleep for periods of apnea and lowered blood oxygen saturation).

The physician must differentiate sleep apnea syndrome from seizure disorder, narcolepsy, or psychiatric depression. A hemoglobin level is obtained, and thyroid function tests are performed. Several clinical diagnostic predictive formulas are being studied.³³² Diagnosis criteria for children have not been standardized, although it is recognized that they should be different from adult criteria.³⁵³ The primary symptom in children is hyperactivity, not daytime somnolence.

TREATMENT. Obstructive and mixed types of sleep apnea syndrome can be treated. Since many clients with sleep apnea are overweight, weight loss is recommended. Weight loss may be curative, but only a small percentage of people maintain their weight loss and symptoms return with weight gain. Therefore alternative interventions have been developed, and the most common treatment for OSA is CPAP used during sleep. The positive pressure from the CPAP pumps open the airway and prevents it from obstructing, but this treatment technique may not be tolerated by some people and adherence to its use is only about 40%.²⁶⁴ Adherence level is demonstrated in the first 2 weeks, but there may be increased compliance after 2 years because the perceived benefits outweigh the barriers such as noise and discomfort.^{151,406}

BiPAP and automatic self-adjusting CPAP have also been used, but their effectiveness is not known at this time.²³² One study has evaluated adherence and effectiveness of CPAP in children. CPAP was effective in improving oxygen saturation and the average nightly use was 5.3 hours (a time markedly overestimated by parents).²⁷⁶

In adults, CPAP is more effective than no treatment and treatment with oral appliances (OA) in reducing apnea, decreasing blood pressure, and improving quality of life.¹⁵⁵ Guidelines for use of CPAP are published.²³² OA may be inserted into the mouth at bedtime and used to hold the jaw forward, thus preventing pharyngeal occlusion. Such devices are regulated by the FDA. There is evidence that OA reduces sleepiness and disordered breathing when compared to control but that CPAP is more effective overall. It is recommended that OA be used for mild sleep disorders or when the person does not tolerate CPAP.²⁶¹

Surgery is recommended if an airway obstruction can be determined as the cause of the sleep apnea. Neuro-

genie causes of sleep apnea are more difficult to control. There is insufficient evidence to draw any conclusions about the effectiveness of medication. Medication has been directed at improving tone in the upper airway, increasing ventilatory drive, reducing REM sleep, and reducing airway resistance or surface tension.³⁹⁰ Alcohol and hypnotic medications should be avoided. In children, tonsillectomy/adenoidectomy has been shown to be effective in a majority of cases.^{353,385}

PROGNOSIS. Evidence indicates that OSA may be associated with increased long-term cardiovascular and neurophysiologic morbidity. Cardiac and vascular morbidity may include systemic hypertension, cardiac arrhythmias, pulmonary hypertension, cor pulmonale, left ventricular dysfunction, stroke, and sudden death. Recognition and appropriate treatment of OSA and related disorders will often significantly enhance the client's quality of life, overall health, productivity, and safety on the highway and job.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-10

Sleep-Disordered Breathing: Apnea

PREFERRED PRACTICE PATTERNS

6A: Primary Prevention/Risk Reduction for Cardiovascular/Pulmonary Disorders (apnea is a risk factor for cardiovascular involvement)

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

There is very little research on the effects of pulmonary rehabilitation in persons with sleep apnea. Two studies demonstrated positive responses to exercise training, although exercise alone was not an adequate intervention.³²³ One small study showed that physical activity level was better correlated with subjective measures of well-being than severity of apnea disorder.²⁰⁴

Pulmonary rehabilitation may be an effective adjunct intervention to improve quality of life and cardiovascular fitness and to assist with weight loss. Because of the possible cardiovascular complications associated with clients who have OSA, vital signs should be monitored before, during, and after submaximal or maximal exercise. The client should not be left in the supine position for prolonged periods of time, even while awake.

There are some reports of sleep apnea in association with cervical lesions (e.g., osteophytes caused by diffuse idiopathic skeletal hyperostosis [DISH]),²⁶⁸ as well as after anterior cervical spine fusion.¹⁸² Individuals with rheumatoid arthritis complicated with temporomandibular joint destruction and cervical involvement can also develop OSA.³³⁰ Physical therapy may be explored in developing treatment protocols when the musculoskeletal structures of the mandible contribute to the problem.

Restrictive Lung Disease

Overview

Restrictive lung disorders are a major category of pulmonary problems, including any condition that reduces the lung volume and decreases compliance. Pulmonary function tests are characterized by a decrease in total lung capacity. There are many causes of restrictive lung diseases that are covered in other sections of this chapter or book. More than 100 identified interstitial lung diseases can cause restrictive lung disease.

Extrapulmonary causes may include neurologic or neuromuscular disorders (e.g., head or spinal cord injury, amyotrophic lateral sclerosis (ALS), myasthenia gravis, Guillain-Barre syndrome, muscular dystrophy, or poliomyelitis), musculoskeletal disorders (e.g., ankylosing spondylitis, kyphosis or scoliosis, or chest wall injury or deformity), postsurgical conditions, particularly involving the abdomen or thorax, and obesity.

Clinical Manifestations

Clinical presentation varies according to the cause of the restrictive disorder. Generally, clients with restrictive lung disease exhibit a rapid, shallow respiratory pattern. Chronic tachypnea (fast rate) occurs in an effort to overcome the effects of reduced lung volume and compliance.

Exertional dyspnea progresses to dyspnea at rest because of the loss of inspiratory reserves. As the disease progresses, respiratory muscle fatigue may occur, leading to inadequate alveolar ventilation and carbon dioxide retention. Hypoxemia is a common finding, especially in the later stages of restrictive disease.

MEDICAL MANAGEMENT

TREATMENT AND PROGNOSIS. The management of restrictive lung disease is based in part on the underlying cause. Treatment goals are oriented toward adequate oxygenation, maintaining an airway, and obtaining maximal function. For example, persons with spinal deformities may be helped with corrective surgery and obese persons may experience improved breathing after weight loss.

Corticosteroids may help control inflammation and reduce further impairment, but previously damaged alveolocapillary units cannot be regenerated or replaced. Some clients with end-stage disease may be candidates for lung transplantation. Most restrictive lung diseases are not reversible, and the disease progresses to include pulmonary hypertension, cor pulmonale, severe hypoxia, and eventual ventilatory or cardiac failure.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-11

Restrictive Lung Disease

PREFERRED PRACTICE PATTERNS

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure

Continued.

Exercise testing (6-minute walk tests or other submaximal exercise evaluation) plays an important role in determining the extent of the disease and assessing outcomes. Many residual affects of pulmonary pathology are neuromuscular in nature and can be addressed by appropriate physical therapy.²⁸³

A primary problem for clients with restrictive lung disease secondary to generalized weakness and neuromuscular disease is ineffective cough. Airway clearance techniques to facilitate cough and effective dislodging of secretions to the central airways may be exhausting for the client. Rest periods must be incorporated in the treatment.

A person with restrictive lung disease will be more adversely affected by the restriction of lung function in the recumbent position, emphasizing the importance of routine positioning for immobile clients and active or active-assisted movements whenever possible. Extrapulmonary causes of restriction are most amenable to physical therapy intervention. Consider manual therapy for improving chest wall compliance with injury or after surgery, as well as flexibility exercises.

Pulmonary Fibrosis

Definition and Overview

Pulmonary fibrosis (also known as interstitial lung disease) is a general term that refers to a variety of disorders in which ongoing epithelial damage or chronic inflammation of lung tissue leads to progressive scarring (fibrosis) of the lungs, predominantly fibroblasts and small blood vessels that progressively remove and replace normal tissue.³²²

Etiologic and Risk Factors

Two-thirds of cases of pulmonary fibrosis are idiopathic pulmonary fibrosis (IPF), in which the cause is unknown. In the remaining one-third, fibrosis in the lung is caused by healing scar tissue after active disease, such as TB, systemic sclerosis, or adult respiratory distress syndrome (ARDS), or after inhalation of harmful particles such as moldy hay, metal dust, coal dust, or asbestos.

Other risk factors include some infections and connective tissue diseases, such as rheumatoid arthritis or SLE; certain drugs, particularly some chemotherapy agents; and in rare cases, genetic or familial predisposition.

Thoracic radiation (e.g., postmastectomy irradiation of the chest wall and regional lymphatics in clients with breast cancer) may result in pericarditis and pneumonitis, which can progress to pulmonary fibrosis weeks, or even months, after radiation treatments have ended (see the section on Radiation Lung Disease in Chapter 5). In addition, some chemotherapies can cause pulmonary fibrosis.³²⁹

Pathogenesis and Clinical Manifestations

Fibroblast proliferation (fibrosis) irreversibly distorts and shrinks the lung lobe at the alveolar level and causes a marked loss of lung compliance. The lung becomes stiff and difficult to ventilate with decreased diffusing capacity of the alveolocapillary membrane, causing hypoxemia. There does not appear to be an inflammatory process but rather abnormal wound healing in response to multiple, microscopic sites of ongoing alveolar epithelial injury and fibrosis.^{321,322} The course of pulmonary fibrosis varies, with early symptoms such as SOB and a dry cough potentially progressing to further complications.

MEDICAL MANAGEMENT

DIAGNOSIS. Definitive diagnosis of IPF is with surgical biopsy. Clinical assessment, pulmonary function tests, and radiographic studies support the pathologic findings.

TREATMENT AND PROGNOSIS. Although past treatment for IPF has included corticosteroids, there is insufficient evidence to support their use. Because of the more recent hypothesis that repeated lung injury is the cause, antiinflammatory treatment is not warranted. Other types of pulmonary fibrosis may respond to corticosteroids.³⁵⁴

Other approaches to treat IPF include immunomodulatory, immunosuppressive, or antifibrotic agents.¹⁰⁸ Novel approaches target growth factors, angiogenesis (formation of new capillaries), cytokines, apoptosis (programmed cellular death), epithelial regeneration, and oxidative stress.¹⁶ These treatments, alone and in combination, require much further study to determine their effectiveness in slowing or curing this disease.

The clinical course of people with pulmonary fibrosis and rheumatoid arthritis is chronic and progressive. Response to treatment is unpredictable, and the overall prognosis is poor, with median survival time less than 4 years.¹⁶²

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-12

Pulmonary Fibrosis

PREFERRED PRACTICE PATTERN

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure (Deconditioning)

One of the most common late effects of chest irradiation is pulmonary fibrosis, which may not occur for months to years after radiation to the thorax. The total dose of radiation and the size of the treatment portal determine the severity of this condition. The changes in pulmonary function are usually a progressive decline in lung volumes and a decrease in lung compliance and diffusing capacity. As doses increase, the frequency of pulmonary fibrosis increases, but with improved dosage fractionation, most people die from the cancer before these complications develop.

Early identification of IPF may improve morbidity. Identifying and referring anyone who has unusual SOB or progressive decrease in exercise tolerance may help with early diagnosis. Physical therapy intervention depends on clinical presentation following the appropriate preferred practice pattern or patterns and may focus on peripheral conditioning and motor control for more efficient oxygen utilization.

Systemic Sclerosis Lung Disease

Definition

Systemic sclerosis (SS), or scleroderma, is an autoimmune disease of connective tissue characterized by excessive collagen deposition in the skin and internal organs, particularly the kidneys and lungs. This condition is discussed in detail in Chapter 10.

Incidence

Clinically, more than one-half of all people with SS die of pulmonary disease.³⁹⁸ The presence of pulmonary arterial hypertension is a major prognostic factor in mortality. The lungs, as a result of a rich vascular supply and abundant connective tissue, are a frequent target organ (second to the esophagus in visceral involvement). Skin changes generally precede visceral alterations, and lung involvement rarely presents symptoms at first, but pulmonary symptoms develop after an average of 7 years.³⁹

Pathogenesis and Clinical Manifestations

Three pathways produce organ damage. First, inflammation is caused by T cells and cytokines, resulting in alveolitis before fibrosis. Second, severe thickening and obstruction of vessels occurs, resulting in pulmonary hypertension and renal failure. Third, cutaneous fibrosis occurs.³⁹⁷ Immunosuppressive therapy may delay onset of symptoms by up to 4 years.³⁹

Oxidative stress contributes to disease progression by a rapid degeneration of endothelial cell function in SS. Daily episodes of hypoxia-reperfusion injury produce free radicals (see Fig. 6-2) that cause endothelial damage, intimal thickening, and fibrosis along with inactivation of antioxidant enzymes.¹⁴⁵

Recent studies have determined that the balance between fibrotic and inflammatory mediators may be important to developing pathology.¹⁹⁶ Lung biopsy of early lesions shows capillary congestion, hypercellularity of alveolar walls, increased fibrous tissue in the alveolar septa, and interstitial edema with fibrosis. As a result, initial symptoms of dyspnea on exertion and nonproductive cough develop. As fibroblast proliferation and collagen deposition progress, fibrosis of the alveolar wall occurs and the capillaries are obliterated. Clinically, the client demonstrates more severe dyspnea and has a greater risk of deterioration in pulmonary function.

MEDICAL MANAGEMENT

DIAGNOSIS. Traditional tests, such as pulmonary function tests and chest radiographs, are insensitive and not

predictive of outcome. Thin-section CT is very sensitive for early diagnosis of SS lung involvement. Bronchoalveolar lavage and serum markers (surfactant protein D and LK-6) give some indication of the disease process.¹⁹⁶

TREATMENT. Successful treatment of SS pulmonary disease remains an area for further development. Pharmacologic treatment using low-dose prednisone is recommended because of the possible association of high-dose corticosteroids with renal failure in clients with SS. Cyclophosphamide, an antineoplastic alkylating agent, has been shown to be effective in treating alveolitis in people with SS.⁴⁶²

Identifying the cycle of oxidative stress and antioxidant inactivation may result in treatment by supplementation of antioxidants and different kinds of drugs with antioxidant properties.^{145,399} Drugs targeted at pulmonary hypertension are currently being studied, including an endothelin receptor antagonist, a prostacyclin analogue, and a phosphodiesterase type 5 (PDE5) inhibitor.^{148,397,462}

Investigations conclude that lung transplantation is a viable option for carefully selected individuals with scleroderma-related lung disease; survival rates are equivalent to lung transplant recipients with other disorders.²⁸⁴

PROGNOSIS. SS lung disease is unpredictable and may be a mild, prolonged course, but as the pulmonary fibrosis advances and causes pulmonary hypertension, cor pulmonale characterized by peripheral edema may develop, progressing rapidly to respiratory failure and death. Lung disease is the most frequent cause of death from SS. Morbidity and mortality of adults over 75 years were worse than younger adults, in part related to late diagnosis.¹¹⁵

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-13

Systemic Sclerosis Lung Disease

PREFERRED PRACTICE PATTERNS

Patterns may vary, depending on degree of pulmonary involvement and response to treatment. See also Special Implications for the Therapist: Systemic Sclerosis in Chapter 10.

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure (pulmonary fibrosis)

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

The effectiveness of a pulmonary rehabilitation program with SS lung disease remains unknown and warrants research investigation. Therapy implications and interventions should be based on general principles regarding pulmonary involvement and specific clinical presentation.

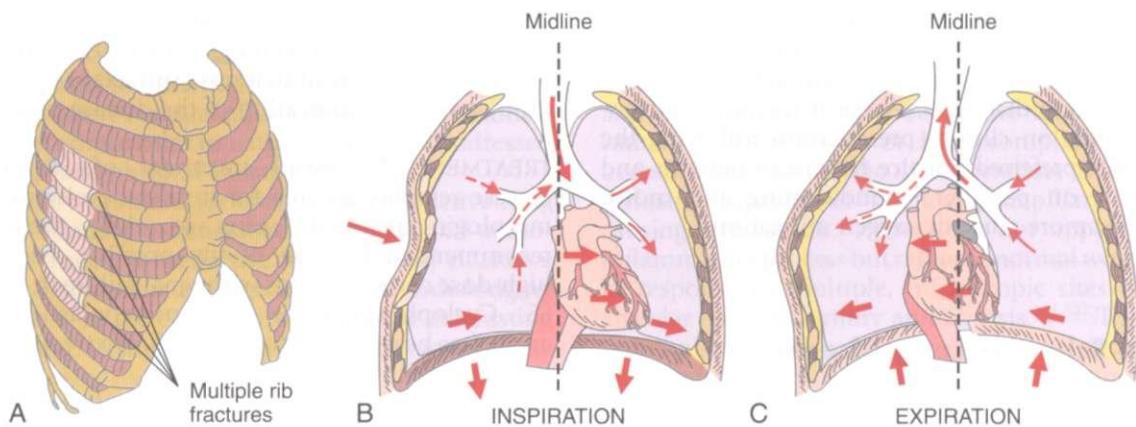


Figure 15-13

Flail chest. Arrows indicate air movement or structural movement. **A**, Flail chest consists of fractured rib segments that are detached (free-floating) from the rest of the chest wall. **B**, On inspiration, the flail segment of ribs is sucked inward. The affected lung and mediastinal structures shift to the unaffected side. This compromises the amount of inspired air in the unaffected lung. **C**, On expiration, the flail segment of ribs bellows outward. The affected lung and mediastinal structures shift to the affected side with the diaphragm elevated on that side (not shown). Some air within the lungs is shunted back and forth between the lungs instead of passing through the upper airway.

Chest Wall Trauma or Lung Injury

Blunt Chest Trauma

Chest or thoracic trauma ranges from superficial wounds such as contusions and abrasions to life-threatening tension pneumothorax. Flail chest occurs as a result of sternum or multiple rib fracture. By definition, a flail chest consists of fractures of two or more adjacent ribs on the same side, and possibly the sternum, with each bone fractured into two or more segments.

The fractured rib segments are detached (free-floating) from the rest of the chest wall. The integrity of the thorax is compromised, and the inspiratory force of the diaphragm causes inward (paradoxical) movement of the fractured ribs. The number of rib fractures is directly correlated with lung-related complications, and the presence of six or more rib fractures significantly increases mortality from nonpulmonary causes.¹³⁴

Early identification improves outcomes, particularly in children in whom chest trauma is the second leading cause of death.³⁷³ Complex soft tissue injury can occur in the absence of chest wall fractures.³ Cough-induced rib fractures occur primarily in women and can occur in persons with normal bone density.¹³⁵ These fractures are typically lateral, in the middle ribs, and do not cause flail chest.

Clinical Manifestations

It is common for a fractured rib end to tear the pleura and lung surface, thereby producing hemopneumothorax. This causes the lung to collapse from the loss of negative pressure. Fractured ribs can also lacerate abdominal organs, the brachial plexus, and blood vessels.

In flail chest, the paradoxical chest motion impairs movement of gas in and out of the lungs (Fig. 15-13), promotes atelectasis, and impairs pulmonary drainage. Other clinical manifestations of flail chest include excruciating pain, severe dyspnea, hypoventilation, cyanosis,

and hypoxemia, leading to respiratory failure without the appropriate intervention.

MEDICAL MANAGEMENT

DIAGNOSIS. In a retrospective study of 492 adults, the combination of pain with palpation and hypoxia predicted 100% of all significant acute intrathoracic injuries seen on radiographs.³⁶⁰ Because blunt trauma may also involve significant soft tissue injury, multidetector CT is recommended to more accurately determine the extent of injury.³⁰¹

Initial treatment follows the ABCs of emergency treatment (airway, breathing, and circulation) to treat the pneumothorax, thereby enabling the person to breathe deeply and to effectively clear secretions. A chest tube removes both air and blood in the pleural cavity after chest trauma and regains the negative pressure in the pleural space so the lungs can remain inflated. The tube is usually positioned in the sixth intercostal space in the posterior axillary line. CPAP or PEEP may be used to enhance lung expansion.²⁸

TREATMENT. Treatment may require internal fixation by controlled mechanical ventilation until the chest wall has stabilized, which may take 14 to 21 days or more. Ventilators are able to monitor pressure, flow, and volume so treatment can be prescribed and modified for each person.⁴³

Whenever pulmonary function is adequate, intubation is avoided to help reduce infection, the most common complication associated with morbidity and mortality in clients with flail chest. Pharmacologic treatment may include muscle relaxants or musculoskeletal paralyzing agents (e.g., pancuronium bromide) to reduce the risk of separation of the healing costochondral junctions.

Older adults are more likely to have comorbid conditions and less likely to tolerate traumatic respiratory compromise. Older adults have a significantly higher rate of

chest injuries sustained in motor vehicle accidents.²⁵⁹ Age and its effects on the body are the strongest predictor of outcome with flail chest, and increasing age is associated with increased complications and mortality.⁸

Ventilator-Induced Injury

Damaged caused by ventilator use is indistinguishable from the damage caused by ARDS.¹⁵⁶ In one study, 24% of the people with no acute lung injury developed acute lung injury from mechanical ventilation within 5 days.¹⁴⁶ Care must be taken to adjust tidal volume using height and gender. Using a prone position may decrease the lung stress while on ventilation.²⁹⁶

Sternal fractures associated with clinically silent myocardial contusion are best visualized on chest CT, but scapular fractures are often overlooked when only supine chest radiographs are performed. The therapist may recognize a suspicious clinical presentation (e.g., loss of scapular-humeral motion, symptoms out of proportion to the injury, or development of previously undocumented large hematomas) suggesting the need for more definitive medical diagnosis. In the case of all fractures, once the fracture is healed the therapist may become involved in restoration of movement and strength.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-14

Chest Wall or Lung Injury

PREFERRED PRACTICE PATTERN

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure (pulmonary insufficiency secondary to injury; pneumothorax; hemothorax; pulmonary collapse)

See also the section on Pneumothorax and Special Implications for the Therapist: Pneumothorax in this chapter.

The emergency room therapist is the most likely therapist to evaluate and treat someone with a flail chest although this varies by geographic region. Transcutaneous electrical nerve stimulators (TENS) have been shown to be more effective than NSAIDs for controlling pain associated with rib fractures.³²⁶ This is important because pain can further compromise pulmonary function.²³⁶

Once the person has been stabilized and moved to the acute care setting, therapists may come into contact during the recovery period. Manual techniques for secretion removal may be used, but the presence of a lung contusion directs medical intervention more toward drainage or aspiration of any blood pooled in the area.

Airway clearance techniques may have a role in facilitating chest tube drainage but must be used carefully in the presence of any rib fractures. Percussion and vibration techniques are contraindicated directly over fractures but can be used over other lung segments. Rib or chest taping and ultrasound over the site of the fracture should not be used. Once the fractures have healed, rib mobilization and soft tissue mobilization for the intercostals may be necessary to restore normal respiratory movements.

It should be noted that airway clearance techniques can cause rib fractures and that infants are particularly vulnerable to rib fractures.³³ Frequent turning and position changes, as well as deep-breathing and coughing exercises, are important. A semi-Fowler's position may help with lung reexpansion necessary to prevent atelectasis. In the case of flail chest from injury, simultaneous cardiac damage may have occurred, necessitating the same care as for a person who has suffered a myocardial infarction (see Special Implications for the Therapist: Myocardial Infarction in Chapter 12).

ENVIRONMENTAL AND OCCUPATIONAL DISEASES

The relationship between occupations and disease has been observed, studied, and documented for many years. An in-depth discussion of this broad topic is included in Chapter 4. This chapter discusses only environmental and occupational diseases related to the lung. Occupational diseases can be divided into three major categories: (1) inorganic dusts (pneumoconioses); (2) organic dusts (hypersensitivity pneumonitis); and (3) fumes, gases, and smoke inhalation. These three categories have pathologic characteristics in common, including involvement of the pulmonary parenchyma with a fibrotic response.

Pneumoconiosis

Overview

Any group of lung diseases resulting from inhalation of particles of industrial substances, particularly inorganic dusts, such as those from iron ore or coal, with permanent deposition of substantial amounts of such particles in the lung, is included in the generic term of *pneumoconiosis* (dusty lungs). Clinically common pneumoconioses include coal workers' pneumoconiosis, silicosis, and asbestosis. Other types of pneumoconiosis include talc, beryllium lung disease (berylliosis), aluminum pneumoconiosis, cadmium workers' disease, and siderosis (inhalation of iron or other metallic particles). Farmers in dry climate regions exposed to respirable dust (inorganic agricultural dusts) during farming activities (e.g., plowing and tilling) and toxic gases (e.g., from animal confinement) may develop chronic bronchitis, hypersensitivity pneumonitis, and pulmonary fibrosis.

Incidence and Etiologic Factors

Obviously occurring in occupational groups, pneumoconiosis is most common among miners, sandblasters, stonemasons, asbestos workers, insulators, and agriculture workers. There is an increasing incidence with age because of cumulative effects of exposure, but overall incidence of diseases caused by mineral dust has declined recently in postindustrial countries. Instead, there is a rise in occupational asthma and illnesses caused by exposure in new office buildings and hospitals.³⁵

Silicosis, formerly called *potters' asthma*, *stonecutters' cough*, *miners' mold*, and *grinders' rot*, is most likely to be contracted in today's industrial jobs involving sandblasting in tunnels, hard-rock mining (extraction and processing of ores), and preparation and use of sand. It can occur in anyone habitually exposed (usually over a period of 10 years) to the dust contained in silica, and any miner is subject to it. Usually, silicosis is associated with extensive or prolonged inhalation of free silica (silicon dioxide) particles in the crystalline form of quartz.

Risk Factors

Higher-risk workplaces are those with obvious dust, smoke, or vapor or those in which there is spraying, painting, or drying of coated surfaces. Heavier exposure occurs when there is friction, grinding, heat, or blasting; when very small particles are generated; and in enclosed spaces.

Not all clients exposed to occupational inhalants will develop lung disease. Harmful effects depend on the (1) type of exposure, (2) duration and intensity of exposure, (3) presence of underlying pulmonary disease, (4) smoking history, and (5) particle size and water solubility of the inhalant. The larger the particle, the lower the probability of its reaching the lower respiratory tract; highly water-soluble inhalants tend to dissolve and react in the upper respiratory tract, whereas poorly soluble substances may travel as far as the alveoli.

The risk of lung cancer in those who both smoke and are exposed to asbestos is increased in a multiplicative way.⁴⁴ Exposure to significant amounts of asbestos is most common when asbestos materials are disturbed during renovation, repair, or demolition of older buildings containing asbestos materials.

Exposure while washing clothes soiled with these toxic substances has caused mesothelioma (malignancy associated with asbestos exposure) and berylliosis (beryllium lung disease associated with exposure to beryllium used in the manufacture of fluorescent lamps before 1950). Beryllium is used today as a metal in structural materials employed in aerospace industries, in the manufacture of industrial ceramics, and in atomic reactors, so exposure is still possible.

Pathogenesis

Dust particles (indestructible mineral fibers) that are not filtered out by the nasociliary mechanism or mucociliary escalator may be deposited anywhere in the respiratory tract and lungs, especially the small airways and alveoli. Each disease has its own pathogenesis, but in general the most dangerous dust particles measure 2 pm or less and are deposited in the smallest bronchioles and the acini (see Fig. 15-2). The particles are ingested by alveolar macrophages, and most of the phagocytosed particles ascend to the mucociliary lining and are expectorated or swallowed. Some migrate into the interstitium of the lung and then into the lymphatics. These indestructible mineral fibers can actually pierce the lung cells. In response to the continued presence of these fibers and to the cell damage, activated macrophages secrete fibroblast-stimulating factor, which in turn mediates excessive fibrosis (i.e., the thickening and scarring of lung tissue that occur around the mineral fiber).

In *coal workers' pneumoconiosis*, ingestion of inhaled coal dust by alveolar macrophages leads to the formation of coal macules, which appear on the radiograph as diffuse small opacities (or white areas) in the upper lung. Anthracite or hard coal is associated with a higher incidence of black lung than is bituminous or soft coal.

In the pathogenesis of silicosis, groups of silicon hydroxide on the surface of the particles form hydrogen bonds with phospholipids and proteins, an interaction that is presumed to damage cellular membranes and thereby kill the macrophage. The dead macrophages release free silica particles and fibrogenic factors. The released silica is then reingested by macrophages, and the process is amplified.

Between 10 and 40 years after initial exposure to silica, small rounded opacities called *silicotic nodules* form throughout the lung. These fibrotic nodules scar the lungs and make them receptive to further complications (e.g., TB, bronchitis, or emphysema).

Asbestosis is characterized by inhalation of asbestos fibers, a fibrous magnesium and calcium silicate non-burning compound used in roofing materials, insulation for electric circuits, brake linings, and many other products that must be fire-resistant. As with the other pneumoconioses, asbestos particles are engulfed by macrophages. Once activated, macrophages then release inflammatory mediators resulting in nodular interstitial fibrosis that can be seen on radiographs along with thickened pleura.

After an interval of 10 to 20 years between exposure and further complications, calcified pleural plaques on the dome of the diaphragm or lateral chest wall develop. The lower portions of the lungs are more often involved than the upper portions in asbestosis.

How asbestos causes mesothelioma is unclear; the formation of oxygen free radicals by macrophages can be a cause of chromosomal damage, or there may be a growth factor that governs individual susceptibility to mineral fiber-induced mesothelioma. Other mechanisms of oncogenesis have been proposed but remain unconfirmed.^{76,353}

Clinical Manifestations

Symptoms of pneumoconioses from dust exposure include progressive dyspnea, chest pain, chronic cough, and expectoration of mucus containing the offending particles. In rare cases, rheumatoid arthritis coexisting primarily with coal workers' pneumoconiosis but also with silicosis and asbestosis causes Caplan's syndrome, a condition characterized by the presence of rheumatoid nodules in the periphery of the lung. Long-term exposure to acid and other substances produces ulceration and perforation of the septum, whereas nickel and certain wood dusts cause nasal carcinoma.

Work-related asthma can be an exacerbation of asthma that was previously subclinical or in remission (work-aggravated asthma), a new onset of asthma caused by a sensitizing exposure (asthma with latency), or asthma that results from a single heavy exposure to a potent respiratory irritant (referred to as asthma without latency, irritant asthma, or reactive airways dysfunction syn-

drome). Symptoms are as discussed in the section on Asthma in this chapter.

Simple silicosis is usually asymptomatic and has no effect on routine pulmonary function tests. As the disease progresses, mucus tinged with blood, loss of appetite, chest pain, and general weakness may occur. In complicated silicosis, dyspnea and obstructive and restrictive lung dysfunction occur.

Asbestosis is characterized by dyspnea, inspiratory crackles (on auscultation), and sometimes clubbing and cyanosis. As in the case of the other pneumoconioses, the simple or uncomplicated form of coal workers' pneumoconiosis is uncommon, but the chronic form is often associated with chronic bronchitis and infections.

MEDICAL MANAGEMENT

PREVENTION. Prevention is the first line of defense against occupational diseases. Workplace-based education, pre-employment screening, yearly physical examinations, surveillance and exposure reduction, and elimination of the pathogen are essential components of a strategy to prevent occupational lung disorders. Precautions, such as the use of facemasks, protective clothing, and proper ventilation, are essential. Regular chest films are recommended for all workers exposed to silica as a means of early detection.

In 1971, asbestos became the first material to be regulated by the U.S. Occupational Safety and Health Administration (OSHA). The Environmental Protection Agency (EPA) has classified asbestos as a Group A human carcinogen, causing both lung cancer and mesothelioma. It is still unclear if lung cancer can occur as result of exposure to asbestos without the presence of asbestosis.¹⁹⁴ The EPA maintains the Integrated Risk Information System database on health effects of exposure to various substances. This agency also controls the National Emission Standards for Hazardous Air Pollutants (<http://www.epa.gov>).

DIAGNOSIS. Identifying a workplace-related cause of disease is important because it can lead to cure and to prevention for others. The recognition of occupational causes can be difficult because of the latency period, delayed responses that occur at home either after work or years after exposure. Diagnosis is by history of exposure (which may be minimal with asbestosis and far removed in time from the onset of disease; the person may even be unaware of the exposure), sputum cytology, lung biopsy, chest film showing nodular or interstitial fibrosis, and pulmonary function studies.

Other pulmonary imaging techniques used in conjunction with the initial chest radiograph include conventional CT, high-resolution CT, and gallium scintigraphy. High-resolution CT scanning is the best imaging method to differentiate different origins of pneumoconiosis as presentation varies with the stimulus (silica, coal dust, iron dust, or asbestos). Magnetic resonance imaging (MRI) is helpful to distinguish progressive fibrosis from lung cancer^{87,275} (see the section on Diagnosis under Lung Cancer in this chapter). Imaging alone is inadequate to make most diagnoses; clinical presentation of symptoms

and lung function are also important.³⁶³ Genetic susceptibility may be associated with beryllium-induced disease and may play a role in mediating other types of pneumoconioses.¹³⁶

TREATMENT. There is no standard treatment for these diseases. The dust deposits are permanent so treatment is directed toward relief of symptoms. Corticosteroids may produce some improvement in silicosis. Although there is no cure for any of the pneumoconioses, the complications of chronic bronchitis, pulmonary hypertension, and cor pulmonale must be treated. When lung neoplasm occurs, surgical removal and therapeutic modalities, such as radiotherapy or chemotherapy, may be employed.

PROGNOSIS. The devastating feature of pneumoconioses is that there may be no obvious symptoms until the disease is in an advanced state. Once fully developed, prognosis is poor for most occupational lung diseases, with progressive and disabling results. Simple silicosis is not ordinarily associated with significant respiratory dysfunction unless complicated by emphysema and chronic bronchitis from cigarette smoking.

Although now uncommon, acute silicosis resulting from heavy exposure to silica rarely responds to treatment and progresses rapidly over a few years when it occurs. The increased incidence of TB among people with silicosis presents an additional negative factor to the prognosis.

Exposure to asbestos, radon, silica, chromium, cadmium, nickel, arsenic, and beryllium may result in neoplasm. Crystalline silica is a known human carcinogen, but this link is not defined and may be overestimated.^{253,339} Both bronchogenic carcinoma and mesotheliomas of the pleura and peritoneum have been linked to asbestos. The exposure typically occurs 20 years before the development of bronchogenic carcinoma and approximately 30 to 40 years before the appearance of mesothelioma. The disease culminates in the sixth decade, with few cases occurring before age 40 years. Although progress has been made, mesothelioma still has a 5-year survival rate of only 9%. New chemotherapy drugs have increased the life expectancy but are not curative.

Coal workers' pneumoconiosis was once thought to cause severe disability, but it is now clear that black lung causes minor impairment of pulmonary function at its worst. When coal miners have severe air-flow obstruction, it is usually due to smoking.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-15

Pneumoconioses

PREFERRED PRACTICE PATTERNS

Other pulmonary pathologic conditions frequently occur in conjunction with pneumoconioses. Preferred practice patterns for chronic bronchitis, cor pulmonale, lung cancer, emphysema, or tuberculosis may also apply.

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

Continued.

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure

New materials are being introduced into the workplace at a faster rate than their potential toxicities can be evaluated despite the fact that many have a pathologic effect on the pulmonary system. The possibility of occupational lung disease should be considered whenever a working or retired person has unexplained respiratory illness.

The minute V/V_{O_2} ratio during exercise is more predictive of clinical dyspnea than airway obstruction in pneumoconioses. This ratio documents the degree of V/Q mismatch and can be useful for therapists in prescribing exercise programs.³³

Steam inhalation and airway clearance techniques, such as controlled coughing and segmental bronchial drainage with chest percussion and vibration, help clear secretions. Exercise tolerance must be increased slowly over a long period beginning with increasing regular activities of daily living. Daily activities should be planned carefully to conserve energy (see Box 9-8), to decrease the WOB, and to afford frequent rest periods.

Graded progression from increasing tolerance for daily activities to a conditioning program may precede or replace an aggressive exercise program. In severe cases, oxygen may be necessary for any increase in activity level or exercise and the person may not progress beyond self-care skills.

Hypersensitivity Pneumonitis

Exposure to organic dusts may result in hypersensitivity pneumonitis, also called extrinsic allergic alveolitis. The alveoli and distal airways are most often involved as a result of inhalation of organic dusts and active chemicals. Most of the diseases are named according to the specific antigen or occupation and involve organic materials such as molds (e.g., mushroom compost, moldy hay, sugar cane, or logs left unprotected from moisture), fungal spores (e.g., stagnant water in air conditioners and central heating units), plant fibers or wood dust (particularly redwood and maple and cotton), cork dust, coffee beans, bird feathers, and hydroxyurea (cytotoxic agent).

Gram-bacterial endotoxins may be more to blame than dust in causing pneumonitis in cotton textile workers.⁴⁴⁰ Mycobacteria have also been shown to be responsible for hypersensitivity pneumonitis in industrial metal grinding and in "hot tub lung."^{7,187}

Regardless of the specific antigen involved in the pathogenesis of hypersensitivity pneumonitis, the pathologic alterations in the lung are similar. A combination of immune complex-mediated and T cell-mediated hypersensitivity reactions occurs, although the exact mechanism of these processes is still unknown.

Host factors, such as cigarette smoking and the presence of some human leukocyte antigen (HLA) proteins, also play an important role in the development or suppression of the disease. Most characteristic is the presence of scattered, poorly formed granulomas that contain

foreign body giant cells. Mild fibrosis may occur, usually in the alveolar walls.

The diagnosis of hypersensitivity pneumonitis of an organic origin is made by history of exposure, pulmonary function studies, inflammatory mediators in sputum, and clinical manifestations, which commonly include abrupt onset of dyspnea, fever, chills, and a nonproductive cough.

Initially, symptoms may be reversed by removing the worker from the exposure (the only adequate treatment), modifying the materials-handling process, or using protective clothing and masks. The symptoms typically remit within 24 to 48 hours but return on reexposure and with time and in some people, may become chronic.

Hypersensitivity pneumonitis may present as acute, subacute, or chronic pulmonary disease depending on the frequency and intensity of exposure to the antigen. The prognosis is poor with repeated exposure to these organic dusts, resulting in nonreversible interstitial fibrosis and other adverse respiratory effects.

Noxious Gases, Fumes, and Smoke Inhalation

Exposure to toxic gases and fumes is an increasing problem in modern industrial society. Any time oxygen in the air is replaced by another toxic or nontoxic agent, asphyxia (deficient blood oxygen and increased carbon dioxide in blood and tissues) occurs. Such is the case when products manufactured from synthetic compounds are heated at high temperatures, releasing fumes. For example, workers who use heating elements to seal meat in plastic wrappers and workers involved in the manufacture of plastics and packaging materials made of polyvinyl chlorides are exposed to these fumes. Workers exposed to the artificial butter flavoring for popcorn, diacetyl, have developed significant respiratory obstruction.²⁵¹

The most common mechanism of injury is local irritation, the specific type and extent depending on the type and concentration of gas and the duration of exposure. For example, highly soluble gases, such as ammonia, rapidly injure the mucous membranes of the eye and upper airway, causing an intense burning pain in the eyes, nose, and throat. Insoluble gases, such as nitrogen dioxide, encountered by farmers cause diffuse lung injury.

Metal fume fever is a systemic response to inhalation of certain metal dusts and fumes such as zinc oxide used in galvanizing iron, the manufacture of brass, and chrome and copper plating. Symptoms include fever and chills, cough, dyspnea, thirst, metallic taste, salivation, myalgias, headache, and malaise. Welding fumes create exposure to multiple hazardous agents and cause varied respiratory and systemic pathology.²⁹⁷ *Polymer fume fever*, associated with heating of polymers, may cause similar symptoms. With brief exposures, the symptoms associated with these two syndromes are self-limiting, but prolonged exposure results in chronic cough, hemoptysis, and impairment of pulmonary function associated with a wide range of lung pathologic conditions.

Chemical pneumonitis can result from exposure to toxic fumes. The acute reaction may produce diffuse lung

injury characterized by air space disease typical of pulmonary edema. In its chronic form, bronchiolitis obliterans develops.

Smoke inhalation injury produces direct mucosal injury secondary to hot gases, tissue anoxia caused by combustion products, and asphyxia as oxygen is consumed by fire. Thermal injury seen in the upper airway is characterized by edema and obstruction. Incomplete combustion of industrial compounds produces ammonia, acrolein, sulfur dioxide, and other substances in today's fires.

Environmental tobacco smoke (ETS), or exposure to secondhand smoke among nonsmokers, is widespread. Home and workplace environments are major sources of exposure. A total of 15 million children are estimated to be exposed to secondhand smoke in their homes annually. ETS increases the risk of heart disease and respiratory infections in children, increases the risk of lung cancer by a factor of 2 to 3, and is responsible for an estimated 3000 cancer deaths of adult nonsmokers and 2300 deaths from sudden infant death syndrome (SIDS) annually.³⁴⁴

Infants born to women exposed to ETS during pregnancy have an increased chance of decreased birth weight and intrauterine growth retardation.¹⁹⁵ Prenatal exposure to mainstream smoke from the mother and even to ETS from the mother has been shown to change fetal lung development and cause air-flow obstruction, promote airway hyperresponsiveness and early development of asthma and allergy, and double the odds of future attention deficit hyperactivity disorder.^{214,256}

Newborns, infants, and children under the age of 2 years are at high risk for cardiovascular effects if they are exposed to household ETS during this time. Endothelial cells of the blood vessels damaged as a result of exposure to passive smoking can be measured during the first decade of life. ETS over a period of more than 10 years changes the intima/media ratio by enhancing the thickness of the vessel wall. Other effects of involuntary smoking among children may include middle ear disease, upper and lower respiratory infections, and asthma.^{215,416}

ETS is associated with rhinitis symptoms of runny nose and nasal congestion in some people and is associated with decreased flow in the airways, bronchial hyperresponsiveness, and increased respiratory infections.¹⁶⁴ Other symptoms following exposure to secondhand tobacco smoke may include headache, chest discomfort or tightness, and cough. See also the section on Lung Cancer in this chapter.

NEAR DROWNING

Definition

Near drowning refers to surviving (24 hours or longer) the physiologic effects of hypoxemia and acidosis that result from submersion in fluid. Near drowning occurs in three forms: (1) dry drowning, inhalation of little or no fluid with minimal lung injury because of laryngeal spasm (10% to 15% of cases); (2) wet drowning, aspiration of fluid with asphyxia or secondary changes caused by aspiration (85%); and (3) recurrence of respiratory

distress secondary to aspiration pneumonia or pulmonary edema within 1 to 2 days after a near-drowning incident. Recovery is rapid if respiration and circulation are restored before permanent neurologic damage occurs. Death may occur from asphyxia secondary to reflex laryngospasm and glottic closure.

Incidence and Risk Factors

Unintentional drowning is the second leading cause of death by injury in those under 15 years of age and the third leading cause of accidental death among all age groups. Nearly 80% of drowning victims are males; other risk factors include epilepsy, mental retardation, heart attack, head or spinal cord injury at the time of the accident, failure to use personal flotation devices, increased use of hot tubs and spas, and lack of proper swimming training or overestimation of endurance by those who can swim. Alcohol consumption while swimming or boating is involved in about 25% to 50% of adolescent and adult deaths associated with water recreation and is a major contributing factor in up to 50% of drownings among adolescent boys.⁸⁰

Pathogenesis

For every child under 15 years who drowns, 5 require medical care for near-drowning injuries. The complications of near drowning fall into two categories: the effects of prolonged anoxia on the brain and kidney, which as end organs may experience complications that are irreversible (determining the final prognosis), and acute lung injury from aspiration of fluids. When aspiration accompanies drowning, severe pulmonary injury often occurs, resulting in persistent arterial hypoxia and metabolic acidosis even after ventilation has been restored.

In the past, a distinction was made between the effects of saltwater and freshwater drowning (e.g., cardiovascular function and changes in blood volume and serum electrolyte concentrations), but it is now known that hypoxia is the most important determinant of survival in human near drowning, regardless of the type of water involved.

Regardless of the amount of water aspirated, the duration of submersion and the water temperature determine the pathologic events. Hypoxia results in global cell damage; different cells tolerate variable lengths of anoxia. Neurons, especially cerebral cells, sustain irreversible damage after 4 to 6 minutes of submersion. The heart and lungs can survive up to 30 minutes.

The extent of CNS injury tends to correlate with the duration of hypoxia, but hypothermia accompanying the incident is associated with changes in neurotransmitter release (glutamate, dopamine) and may reduce the cerebral oxygen requirements and help reduce CNS injury. For a detailed review of cold-water submersion, its mechanisms, and its effects the reader is referred to other sources.^{153,189}

Clinical Manifestations

The clinical features in near drowning are variable, and the person may be unconscious, semiconscious, or awake but apprehensive. Pulmonary and neurologic symptoms predominate, with cough, tachypnea, and possible devel-

opment of ARDS (see discussion in this chapter) with progressive respiratory failure.

Other pulmonary complications include pulmonary edema, bacterial pneumonia, pneumothorax, or pneumomediastinum secondary to resuscitation efforts. Fever occurs in the presence of aspiration during the first 24 hours but can occur later in the presence of infection.

Early neurologic manifestations include seizures, especially during resuscitative measures, and altered mental status, including agitation, combativeness, or coma. Speech, motor, or visual abnormalities may occur, improve gradually, and resolve over several months.

MEDICAL MANAGEMENT

PREVENTION. Prevention of drowning and near-drowning events is a vital part of education. The CDC recommends mandating and enforcing legal limits for blood alcohol levels during water recreation activities, public education about the danger of combining alcohol (and other substances) with water recreation, restricting the sale of alcohol at water recreation facilities, and eliminating advertisements that encourage alcohol use during boating. Additional safeguard techniques for prevention of drowning among children are available.⁸⁰

TREATMENT. Improved training in cardiopulmonary resuscitation (CPR) has resulted in survival of the majority of near-drowning victims who live long enough to receive hospital care. Restoration of ventilation and circulation by means of resuscitation at the scene of the accident is the primary goal of treatment to restore oxygen delivery and prevent further hypoxic damage. Other treatment is largely supportive, with antibiotics for pulmonary infection, maintenance of fluid and electrolyte balance, possible transfusion for significant anemia, and management of acute renal failure.

Comatose near-drowning victims frequently have elevated intracranial pressure caused by cerebral edema and loss of cerebrovascular autoregulation. Reduction of cerebral blood flow adds ischemic injury to already damaged brain tissue. In order to reserve cerebral function in such cases, cerebral resuscitation (controlled hyperventilation; deliberate hypothermia; or use of barbiturates, glucocorticoids, and diuretics) may be utilized.

PROGNOSIS. The prognosis depends in large part on the extent and duration of the hypoxic episode. People have survived as long as 70 minutes of immersion with complete recovery, but up to 20% of all near-drowning victims will have permanent sequelae, many of which are ultimately fatal. If laryngospasm is finally overcome and the person aspirates water or if aspiration of vomitus occurs during resuscitative measures, prognosis is worse than without these complications.

Other unfavorable prognostic indicators include first blood pH values below 7, low rectal temperature on admission to the hospital, abnormal electroencephalogram (EEG), deterioration of room air oxygen saturation, and degree of EEG disturbance.

Coincident head trauma or subdural hematoma presents an additional prognostic complication. Neurologic injury is the most serious and least reversible complica-

tion in those persons successfully resuscitated. Little, if anything, has been shown to help, and it carries a grave prognosis.

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-16

Near Drowning

PREFERRED PRACTICE PATTERNS

Other nonpulmonary components associated with near drowning are included in the neuromuscular patterns: 5B, 5C, 5D, and 5I.

7A: Primary Prevention/Risk Reduction for Integumentary Disorders (pressure ulcers)

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

All near-drowning victims should be admitted to a hospital inpatient setting for observation over 24 to 48 hours because of the possibility of delayed drowning syndrome with confusion, substernal pain, and adventitious breath sounds (rales or rhonchi). The therapist is often involved early on in the case management, providing bedside care much the same as for a person with traumatic brain injury or spinal cord injury. It is not uncommon for a near-drowning accident to be associated with either traumatic brain injury or spinal cord injury.

Evaluate cardiopulmonary status, monitor vital signs, and observe respirations. Airway clearance techniques may be necessary. To facilitate breathing, elevate the head of the bed slightly if possible; observe for signs of infection (see Box 8-1); and check for any areas of skin pressure or factors precipitating pressure ulcers. Provide passive or active-assistive exercise according to the person's functional abilities, and progress as quickly as possible given the medical status.

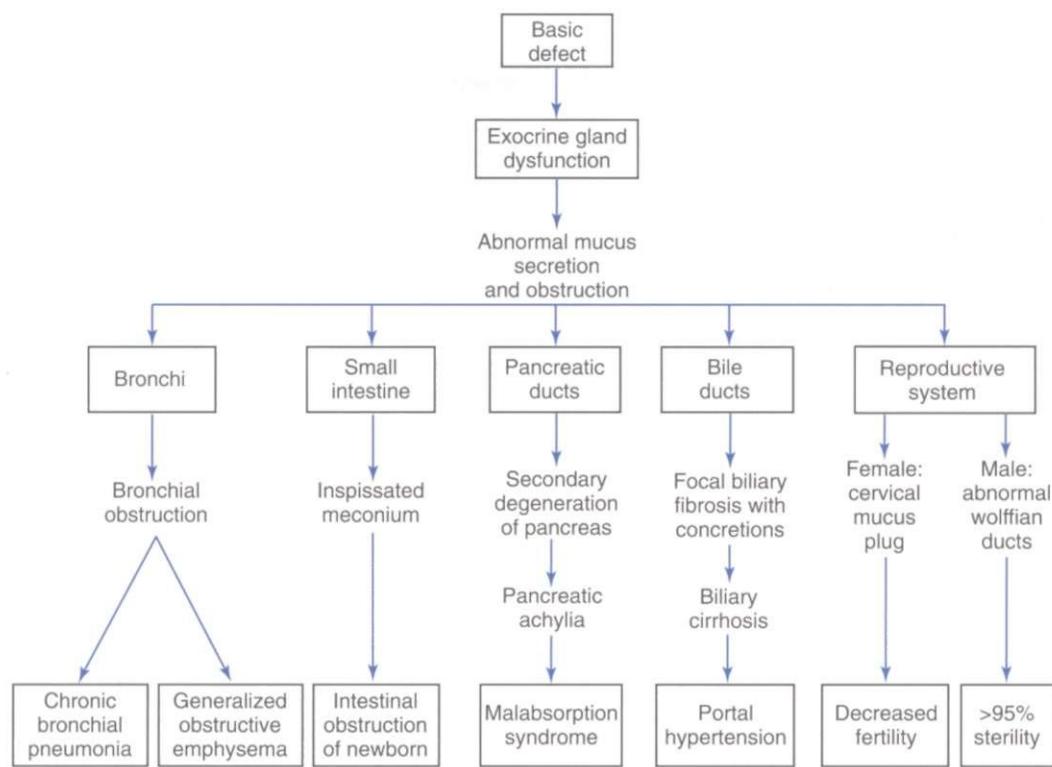
In the rehabilitation setting, large doses of steroids are administered early in the treatment of some cases of spinal cord injury to control cerebral or spinal cord edema. Suppression of the inflammatory reaction in persons receiving large doses of steroids may be so complete as to mask the clinical signs and symptoms of major diseases, perforation of a peptic ulcer, or spread of infection. See also the section on Corticosteroids in Chapter 5.

CONGENITAL DISORDERS

Cystic Fibrosis

Definition and Overview

CF is an inherited disorder of ion transport (sodium and chloride) in the exocrine glands affecting the hepatic, digestive, male reproductive (the vas deferens is functionally disrupted in nearly all cases), and respiratory systems (Fig. 15-14). The basic genetic defect predisposes to

**Figure 15-14**

Various effects of exocrine gland dysfunction in cystic fibrosis. (From Wong DL: *Whaley and Wong's essentials of pediatric nursing*, ed 4, St Louis, 1993, Mosby.)

chronic bacterial airway infections, and almost all persons develop obstructive lung disease associated with chronic infection that leads to progressive loss of pulmonary function.

Incidence

CF is the most common inherited genetic disease in the white population, affecting approximately 30,000 children and young adults (equal gender distribution) in the United States. More than 1000 new cases are diagnosed each year. The disease is inherited as an autosomal recessive trait, meaning that both parents must be carriers so that the child inherits a defective gene from each one. In the United States, 5% of the population, or 12 million people, carry a single copy of the CF gene. Each time two carriers conceive a child, there is a 25% chance (1:4) that the child will have CF, a 50% (1:2) chance that the child will be a carrier, and a 25% chance that the child will be a noncarrier.

Ten percent of new cases are diagnosed in those over 18 years of age.¹⁰⁴ The severity of the disease is strongly correlated with socioeconomic status and access to health care.³⁷⁶

Etiologic Factors

In recent years, there have been major advances in understanding the underlying genetic factors related to this disease. In 1985, the CF gene was located on the long arm of chromosome 7. In 1989, the gene for CF was cloned and abnormalities in the CF transmembrane conductance regulator (CFTR) protein were attributed to CF.

Clones are identical copies of genes used to study the DNA sequence that allows scientists to determine the nature and function of the protein encoded by the gene. Cloning opens up the possibility of gene therapy for a disorder. It should be noted that the CFTR genotype is not a good predictor of disease severity, and a modifier gene has yet to be identified.^{101,105}

In healthy people, this CFTR protein provides a channel by which chloride (a component of salt) can pass in and out of the plasma membrane of many epithelial cells, including those of the kidney, gut, and conducting airways. Clients with CF have a defective gene that normally enables cells to form or regulate that channel.

At least two gating mechanisms of CFTR are now known; one relies on hydrolysis and the second depends on stable adenosine triphosphate (ATP) binding.¹⁸⁸ There are complex relationships between CFTR, the epithelial sodium channel, and mucociliary clearance, which are currently being examined.²⁰⁶ The loss of CFTR function appears to be as important as the defective transport channel.³⁷

Over 300 mutations in the CF gene affecting the CFTR protein have been described, but not all of the mutations have been identified, so mass screening cannot yet identify individuals carrying the gene for CF who would otherwise test negative. New tests are being developed to more reliably detect for mutations.²³²

Inflammation plays a role in lung damage associated with CF unrelated to the genetic defect.¹¹⁰ The role of polymorphism (individual variation) of a gene that regulates protection from lung injury (by producing a sub-

Table 15-10 Respiratory Disease Summary of Differences

Disease	Primary Area Affected	Result
Acute bronchitis	Membrane lining bronchial tubes	Inflammation of lining.
Bronchiectasis	Bronchial tubes (bronchi or air passages)	Bronchial dilation with inflammation.
Pneumonia	Alveoli (air sacs)	Causative agent invades alveoli with resultant outpouring from lung capillaries into air spaces and continued healing process.
Chronic bronchitis	Larger bronchi initially; all airways eventually	Increased mucus production (number and size) causing airway obstruction.
Emphysema	Air spaces beyond terminal bronchioles (alveoli)	Breakdown of alveolar walls; spaces enlarged.
Asthma	Bronchioles (small airways)	Bronchioles obstructed by muscle spasm, swelling of mucosa, thick secretions.
Cystic fibrosis	Bronchioles	Bronchioles become obstructed and obliterated; later larger airways become involved; mucous plugs cling to airway walls, leading to bronchitis, bronchiectasis, atelectasis, pneumonia, or pulmonary abscess.

From Goodman CC, Snyder TE: *Differential diagnosis for the physical therapist*, ed 4, Philadelphia, 2007, Saunders

stance called *glutathione*) has strong association with the severity of CF lung disease.²⁹²

Pathogenesis

Much about the complex pathogenesis of CF is still unknown, but it does appear that this impermeability of epithelial cells to chloride results in (1) dehydrated and increased viscosity of mucous gland secretions, primarily in the lungs, pancreas, intestine, and sweat glands; (2) elevation of sweat electrolytes (sodium chloride); and (3) pancreatic enzyme insufficiency. The dehydration resulting in thick, viscous mucous gland secretions causes the mechanical obstruction responsible for the multiple clinical manifestations of CF.

Bronchial and bronchiolar obstruction by the abnormal mucus predisposes the lung to infection and causes patchy atelectasis with hyperinflation. The disease progresses from mucous plugging and inflammation of small airways (bronchiolitis) to bronchitis, followed by bronchiectasis, pneumonia, fibrosis, and the formation of large cystic dilations that involve all bronchi. A summary of differences among these various respiratory diseases is provided (Table 15-10).

New findings about how specific molecules signal the onset of inflammation and tissue-damaging enzymes and how these chemicals all interact are adding to the knowledge base of CF pathophysiology. For example, in CF lungs, a decreased level of nitric oxide (NO; this is not the same as nitrous oxide [N₂O]), a chemical that decreases inflammation, may be explained by the loss of an enzyme needed to produce NO.

Several laboratories are working to identify how a defect in CFTR causes the loss of NO or the needed enzymes. The inflammatory response against bacteria in the airways of individuals with CF involves the generation of reactive oxygen species (formation of free radicals) leading to further inflammation and tissue damage. Without the necessary NO, a vicious cycle of inflammatory-immune processes and bacteria survival persists. Restoring the balance of oxidants and antioxidants could restore health in the CF lung.

New data on the structure of CFTR, including the size and shape of the channel through which the chloride must pass, how to stabilize the channel and increase the time it stays open, and the life cycle of this protein, are being used to provide scientists with ideas for new treatments.

Other researchers are investigating why the CF lung is so receptive to the onslaught of infection by examining the role of defensin and other bacteria-killing molecules in the CF airway and the inhibition of these antimicrobial peptides by high salt concentrations.

Clinical Manifestations

The consistent finding of abnormally high sodium and chloride concentrations in the sweat is a unique characteristic of CF. Parents frequently observe that their infants taste salty when they kiss them. Almost all clinical manifestations of CF are a result of overproduction of extremely viscous mucus and deficiency of pancreatic enzymes.

A complete list of clinical manifestations by organ and in order of progression is given in Box 15-7. Recurrent pneumothorax, hemoptysis, pulmonary hypertension, and cor pulmonale are serious and life-threatening complications of severe and diffuse CF pulmonary disease.

Pancreas. Approximately 90% of clients have pancreatic insufficiency with thick secretions blocking the pancreatic ducts and causing dilation of the small lobes of the pancreas, degeneration, and eventual progressive fibrosis throughout. The blockage also prevents essential pancreatic enzymes from reaching the duodenum, thus impairing digestion and absorption of nutrients. Clinically, this process results in bulky, frothy (undigested fats because of a lack of amylase and trypsin enzymes), and foul-smelling stools (decomposition of proteins producing compounds such as hydrogen sulfide and ammonia).

As the life expectancy for people with CF has improved, the incidence of glucose intolerance and CF-related diabetes has increased because pancreatic damage can eventually affect the beta-cells. Hyperglycemia may adversely influence nutritional status and weight, pulmonary

Box 15-7**CLINICAL MANIFESTATIONS OF CYSTIC FIBROSIS*****Early Stages***

Persistent coughing
Sputum production
Persistent wheezing
Recurrent pulmonary infection
Excessive appetite, poor weight gain
Salty skin and sweat
Bulky, foul-smelling stools

Pulmonary***Initial***

Wheezy respirations
Dry, nonproductive cough

Progressive Involvement

Increased dyspnea
Decreased exercise tolerance
Paroxysmal cough
Tachypnea
Obstructive emphysema
Patchy areas of atelectasis
Nasal polyps, chronic sinusitis

Advanced Stage

Barrel chest
Kyphosis
Pectus carinatum
Cyanosis
Clubbing (fingers and toes)
Recurrent bronchitis
Recurrent bronchopneumonia
Pneumothorax
Hemoptysis
Right-sided heart failure secondary to pulmonary hypertension

Gastrointestinal

Voracious appetite (early)
Anorexia (late)
Weight loss

Failure to thrive or grow; protein-calorie malnutrition
Distended abdomen
Thin extremities
Sallow (yellowish) skin
Acute gastroesophageal reflux (GERD)
Intussusception

Distal Intestinal Obstruction Syndrome (Meconium Ileus)

Abdominal distention
Colicky, abdominal pain
Vomiting
Failure to pass stools (constipation)
Rapid development of dehydration
Anemia

Liver

Cirrhosis
Portal hypertension

Pancreatic

Large, bulky, loose, frothy, foul-smelling stools (pancreatic enzyme insufficiency)
Fat-soluble vitamin deficiency (vitamins A, D, E, K)
Recurrent pancreatitis
Iron deficiency anemia
Malnutrition
Diabetes mellitus

Genitourinary

Male urogenital abnormalities
Delay in sexual development
Sterility (most males); infertility (some females)

Musculoskeletal

Marked tissue wasting, muscle atrophy
Myalgia
Osteoarthropathy (adult)
Rheumatoid arthritis (adult)
Osteopenia/osteoporosis (adult)

function, and development of late microvascular complications.⁴⁴⁸

Gastrointestinal. The earliest manifestation of CF, *meconium ileus* (sometimes referred to as distal intestinal obstruction syndrome), is present in approximately 10% to 15% of newborns with CF; the small intestine is blocked with thick, puttylike tenacious meconium. Pro-lapse of the rectum is the most common gastrointestinal complication associated with CF, occurring most often in infancy and childhood.

Children of all ages with CF are susceptible to intestinal obstruction from thickened, dried, or impacted stools (inspissated meconium). Advances in investigative techniques have led to increasing reports of Crohn's disease and ischemic bowel disease in persons with CF. Prolonged administration of excessive doses of pancreatic enzymes is associated with the development of fibrosing colonopathy. To avoid this complication, current recommendation for a daily dose of pancreatic enzymes for

most people with CF is below 10,000 units of lipase per kilogram per day.

A new enzyme, TheraCLEC-Total (TCT), has been shown to be promising in improving absorption of fat and nutrients and is ready for type 2 clinical trials.⁵⁰ Poor nutrition and weight loss are common as a result of malabsorption, inadequate oral intake, early satiety, and increased utilization of calories.

Pulmonary. Chronic cough and purulent sputum production are symptomatic of lung involvement. The child is unable to expectorate the mucus because of its increased viscosity. This retained mucus provides an excellent medium for bacterial growth, placing the individual at increased risk for infection. Reduced oxygen-carbon dioxide exchange causes variable degrees of hypoxia, clubbing (see Fig. 15-4), cyanosis, hypercapnia, and resultant acidosis. Chronic pulmonary infection and hyperinflation lead to secondary manifestations of barrel chest, pectus carinatum, and kyphosis.

Box 15-8**SIGNS AND SYMPTOMS OF PULMONARY EXACERBATION IN CYSTIC FIBROSIS**

- Increased cough
- Increased sputum production and/or a change in appearance of sputum
- Fever
- Weight loss
- School or work absenteeism (because of illness)
- Increased respiratory rate and/or WOB
- New findings on chest examination (e.g., wheezing, crackles)
- Decreased exercise tolerance
- Decrease in FEV₁ of 10% or more from baseline value within past 3 months
- Decrease in hemoglobin saturation of 10% or more from baseline value within past 3 months
- New finding(s) on chest x-ray

From Cystic Fibrosis Foundation: *Clinical practice guidelines for cystic fibrosis*, Bethesda, Md, 1997, The Foundation.

WOB, Work of breathing; FEV₁, forced expiratory volume in 1 second.

The most common complication of CF is an exacerbation of pulmonary disease requiring medical and physical therapy intervention. Early warning signs (Box 15-8) must be recognized and treatment initiated (referred to as a "tune-up"), preferably at home but sometimes in the hospital. Respiratory failure is a frequent complication of severe pulmonary disease in persons with CF and is the most common cause of CF-related deaths.

Liver. Liver involvement in CF is much less frequent than both pulmonary and pancreatic diseases, which are present in 80% to 90% of individuals with CF. Liver disease affects only one third of the CF population; however, because of the decreasing mortality from extrahepatic causes, its recognition and management are becoming a relevant clinical issue.^{92a}

Recent observations suggest that clinical expression of liver disease in CF may be influenced by genetic modifiers; their identification is an important issue because it may allow recognition of people at risk for the development of liver disease at the time of diagnosis of CF and early institution of prophylactic strategies.^{92a}

Genitourinary. Genitourinary manifestations are primarily related to reproduction; infertility once thought to be universal in men and common in women may be treated successfully with new techniques for in vitro fertilization. The vas deferens may be absent bilaterally, or if present, it is obstructed so that although sperm production is normal, blockage or fibrosis of the vas deferens prevents release of the sperm into the semen (azoospermia). Women experience decreased fertility because thick mucus in the cervical canal prevents conception. As the disease progresses, there is also an increased incidence of amenorrhea.

Musculoskeletal. Muscle pain is reported and may be alleviated with proper nutrition and exercise, although this is based on anecdotal information and has not been verified in studies. Decreased BMD and bone mineral content are common at all ages in CF, attributed to multifactorial causes (e.g., nutrition, exposure to glucocorti-

coid therapy, gonadal dysfunction, age, body mass, or activity). Spinal consequences of bone loss include excessive kyphosis and neck and back pain. Lung transplant is also associated with increased osteoporosis from long-term immunosuppression.

Hypertrophic pulmonary osteoarthropathy occurs with increasing frequency with increasing age and severity of disease in 2% to 7% of affected individuals. This condition is accompanied by clubbing of the fingers and toes; arthritis; painful periosteal new bone formation (especially over the tibia); and swelling of the wrists, elbows, knees, or ankles. The periostitis is observed radiographically in the diaphysis of the tubular bones and may be a single layer or a solid cloaking of the bone.

Separately and usually without association with other manifestations of CF, attacks of episodic arthritis accompanied by severe joint pain, stiffness, rash, and fever may occur intermittently but repeatedly. Also related to CF are rheumatoid arthritis, spondyloarthropathies, sarcoidosis, and amyloidosis, which are caused by coexistent conditions and drug reactions.⁵¹

MEDICAL MANAGEMENT

DIAGNOSIS. Now that the gene responsible for CF has been identified, prenatal diagnosis and screening of carriers are possible as part of genetic counseling. The tests only detect mutations already observed but account for 70% (those with the DF508 mutation) of all CF carriers. In 2004, the CDC issued a recommendation that all newborns should be screened for CF and currently 18 states and the District of Columbia do routine screening. Pre-pregnancy genetic testing that involves DNA analysis of oocytes is available for couples at risk for having children with CF.

About one-half of all children with CF present in infancy with failure to thrive, respiratory compromise, or both. The age at presentation can vary, and some people are not diagnosed until adulthood. CF is traditionally diagnosed using the sweat test; a positive test occurs when the sodium chloride concentration is greater than 60 mEq/L for anyone younger than 20 years (reference value: 40 mEq/L) and above 80 mEq/L for those over 20 years.^{39b}

Although elevated sweat electrolytes are associated with other conditions, a positive sweat test coupled with the clinical picture usually confirms the diagnosis. The test should be performed at a certified CF center and repeated a second time. Alternatively, CF can be diagnosed by genotype analysis (performed prenatally or postnatally). CF can be diagnosed on DNA alone.

Pancreatic elastase-1 (EL-1), a marker of exocrine pancreatic insufficiency in CF, can be measured in feces. EL-1 is a specific human protease synthesized by the acinar cells of the pancreas and is a reliable test of pancreatic sufficiency over the age of 2 weeks. Fecal elastase has good sensitivity and specificity and predictive values for severe cases of pancreatic insufficiency. This test can be used to rule out the diagnosis of CF, to confirm the need for pancreatic enzymes, and for annual monitoring of pancreatic-sufficient individuals to detect the onset of pancreatic insufficiency. Other tests, including a breath test, are being developed and tested.⁵⁰

Pulmonary function tests are performed in affected individuals from the age of 6 and up to measure and monitor lung function over time (see Table 40-22). These tests are used to classify the severity of baseline lung disease. Almost all measures are based on the flow of air into and out of the lungs in a given period of time.

The two most common lung function measures are FEV₁ (forced expiratory volume in 1 second) and FVC (forced vital capacity). These tests should be repeated two to four times each year for adults to assess the effectiveness of treatment; pulmonary function declines with progressive lung disease.

Diabetes mellitus should be identified early by screening with a glucose tolerance test from the age of 15 years and treated with insulin, dietary management, and exercise from the time of diagnosis of diabetes. Symptoms of CF-related diabetes are often confused with pulmonary infection. Diabetes significantly impacts the course of CF, though the relationship is not clear. The microvascular system should be screened annually for complications.⁹⁶

Several scoring systems have been developed to assess disease severity, measure acute changes, and evaluate appropriateness for lung transplant. The most reliable and useful are the modified Shwachman and modified Huang scores. There is a need for a longitudinal assessment tool to follow individuals with milder CF.¹⁸³

Diagnosis of CF in adulthood is generally due to a milder presentation of the disease and has a more favorable prognosis. Adults with unexplained chronic respiratory infections, bronchiectasis, pancreatitis, or absence of vas deferens should be screened for CF.³¹⁹⁻³⁹⁶

TREATMENT. A multidisciplinary approach must be taken in treating CF toward the goal of promoting a normal life for the individual. The treatment of CF depends on the stage of the disease and which organs are involved. Medical management is oriented toward alleviating symptoms and includes the use of antibiotics, aggressive pulmonary therapy with drugs (mucolytics) to thin mucous secretions, airway clearance techniques, supplemental oxygen, and adequate hydration and enhanced nutrition with pancreatic enzymes administered before or with meals.

Pharmacotherapy. Drug therapy for CF has been primarily directed at prophylaxis and treatment of infections with antibiotics, targeting inflammation, and supplementing digestive enzymes and vitamins. Pharmacotherapy (Table 15-11) to date has included broad-spectrum antimicrobials to protect the respiratory epithelium from damage and aerosolized antibiotics (e.g., tobramycin) that deliver a more concentrated dose directly to the site of infection.

Macrolides, a class of antibiotics, have the added benefit of suppressing inflammatory mediators and interfering with biofilm that is produced by *Pseudomonas aeruginosa*.⁹² Although no randomized control studies have been conducted, a totally implantable venous intravenous access device (TIVAD) for adults is widely used for CF as a means of providing long-term intravenous access for those individuals requiring intermittent antibiotics. The risks of mechanical failure, sepsis, and thrombosis

Table 15-11 Pharmacotherapy for Cystic Fibrosis

Maintenance	Antibiotics, inhaled or oral Antiinflammatory agents Corticosteroids (inhaled or oral) Ibuprofen Bronchodilators Mucolytic expectorants (with airway clearance)
Acute exacerbations	Antibiotics, inhaled or oral

Courtesy Susan Queen, PT, PhD, University of New Mexico, Albuquerque, NM.

have made this device more successful when inserted and cared for at a CF center.^{17,129,235}

Other pharmacologic treatment may include sympathomimetics to control bronchospasm, parasympatholytics to offset smooth muscle constriction and bronchodilation, inhaled antiinflammatory agents to decrease the amount of inflammation in the airways, and mucolytics to thin mucous secretions. Inhaled bronchodilators are effective in individuals who have bronchial hyperresponsiveness.¹⁸⁵

Recombinant human deoxyribonuclease I (rhDNAase I), or Dornase alfa, a mucolytic, is effectively used to reduce sputum viscosity and increase mucociliary clearance.⁴⁰⁹ The use of hypertonic saline, an inexpensive, "low-tech" intervention, has gained in popularity since the hypertonic saline study in Australia. First, a bronchodilator is administered to open the airways, then the saline solution is used to replenish salt depleted from the liquid that lines the airways. This combination of treatment results in fewer flare-ups and faster recovery and is just one of many newer CF treatments.¹²⁶

Hypertonic saline improves airway clearance but is not as effective as DNAase in longer term lung improvement.⁴⁴² Administered 30 minutes before airway clearance treatments, these combined treatment interventions improve the quality of life for people with CF by decreasing their hypoxemia and reducing their dyspnea. The end result is improved sleep patterns, increased activity, and improved nutritional status.

High-dose ibuprofen (the generic name for the drug found in Advil, Motrin, and Nuprin) may be used to slow the deterioration of the lungs by reducing inflammation and breaking the cycle of mucus buildup, infection, and inflammatory destruction. Neutrophils are responsible for much of the inflammatory response and are unresponsive to traditional chemotherapeutic treatment.

New knowledge as to the mechanisms controlling Ca(2+) homeostasis is stimulating novel approaches to stemming the inflammatory response.⁴²¹ Leukotriene-receptor antagonists also have shown the potential to reduce inflammation and improve lung function.³⁷⁸ Investigations into the role of platelets in contributing to the inflammatory response in CF are currently underway.³²⁸

Pharmacologic intervention with microencapsulated pancreatic enzymes is critical when pancreatic involve-

ment is severe. Aggressive nutritional management is needed to ameliorate the effects of malabsorption and the side effects of therapeutic intervention. Calcium supplements are warranted because of the high incidence of osteopenia and fractures.²⁰

Supplemental oxygen may improve exercise endurance and peak performance. Mild hypercapnia (too much carbon dioxide in the blood) can occur with exercise and during sleep. More research is needed to assess the benefits of oxygen therapy.²¹ Oral bile acid therapy, aimed at improving biliary secretion in terms of bile viscosity and bile acid composition, is currently the only available therapeutic approach for CF-associated liver disease.^{92,3}

Gene Therapy. The identification of the mutated CF gene in 1989 was followed by the first phase of gene therapy in 1993 to correct the basic defect in CF cells, rather than relying on treatment of the symptoms. Finding a way to deliver the normal copy of the gene into the lung or intrahepatic biliary epithelial cells with adequate gene expression remains a challenge. Obstacles include vector toxicity and ineffective transgene expression.^{41,7}

It is possible that delivery through an aerosolized technique will incorporate sufficient quantities of the CFTR gene into the cells without toxicity and stimulating an immune response.¹¹⁴ The expectation is that the normal CFTR will reverse the physiologic defect in CF cells.

In the future, current therapeutic measures, such as intravenous antimicrobial treatment, will be improved by the additional delivery of new drugs to the bronchial tree by aerosol. Antibiotics, as well as protease inhibitors delivered by aerosol, should help to prevent damage by infection and inflammation and increase the probability of successful somatic gene therapy in this disease.

Transplantation. Double-lung or heart-lung transplants have been used with children and adults with advanced pulmonary vascular disease and who are severely disabled by dyspnea and hypoxia. In the United States, the United Network for Organ Sharing has addressed perceived inequities in organ distribution by allocating organs by illness severity rather than time on the waiting list. A lung allocation score ranks severity for patients 12 years of age and older for transplantation based on variables, including lung function, oxygen and ventilatory needs, diabetes, weight, and physical performance.^{261a}

Liver transplantation should be offered to anyone with CF and progressive liver failure and/or with life-threatening sequelae of portal hypertension, who also have mild pulmonary involvement that is expected to support long-term survival.^{92a}

Long-term survival has yet to be determined, but improved quality of life has been achieved. The new lungs do not acquire the CF ion-transport abnormalities but are subject to the usual posttransplant complications. CF problems in other organ systems persist and may be worsened by some of the immunosuppressive regimens.⁴⁵⁹

CNS complications occur more frequently in CF transplant recipients than in other lung transplant recipients.¹⁶⁸ Criteria for lung transplant are published, and early referral and continuous monitoring are required to anticipate decline as a result of the long waiting period.¹⁶⁰

PROGNOSIS. Using its innovative CF patient registry, which tracks information on approximately 23,000 clients who receive care through the CF Foundation's Care Center Network, researchers have analyzed the numbers and continue to assess trends in the health status of registered individuals. When first distinguished from celiac disease in 1938, life expectancy with CF was approximately 6 months.¹¹⁰ Data shows that the prognosis has steadily improved over the past 20 years with a gradual increase in longevity; at the time of this publication the median survival had risen to 36.8 years.

More than 50% of children with CF live into adulthood. The new median age of survival is based on 2005 data that includes date of birth, date of death, gender, and date of diagnosis. A detailed CF Foundation Annual Patient Registry Data Report is available.

Improvement in both the length and quality of life for adults with CF is primarily the result of standardization of care, implementation of best practices, continuous multidisciplinary care provided by specialists in CF centers, improved CF therapies, and improved nutrition and pulmonary function, which are two main prognostic factors for improved survival.²⁶⁹

Children whose presenting symptoms were gastrointestinal at diagnosis have a good clinical course; those whose initial symptoms at diagnosis were pulmonary frequently demonstrate subsequent clinical deterioration. Pulmonary failure is still the most common cause of death. Males have a more favorable prognosis than females. New agents and gene therapy may substantially change the morbidity and mortality of this disease with continued improved survival time.

Lung disease is the primary cause of death for 80% for individuals with CF. Lung transplantation is an important therapeutic option for this group, comprising the third largest group of transplant recipients receiving lung transplantation (after people with chronic obstructive lung disease and IPF). The primary goal of transplant has always been to treat individuals with CF with end-stage lung disease for whom medical therapies have failed. The secondary goal has been to improve quality of life.^{261a}

Since the first lung transplant for CF in 1983, survival rates have improved. Refinements in surgical technique, medications, and improved selection criteria have gradually improved postsurgical survival.^{261a} Individuals with CF who are listed for lung transplantation may require mechanical ventilatory support before transplant. Pre-transplant mechanical ventilation increases short-term morbidity and mortality in pediatric clients.^{125a} Besides ventilator dependence, other negative predictive factors for prognosis after transplantation include *Burkholderia cepacia* infection, young age, and arthropathy.^{261a}

The 3-year predicted survival rate after lung transplantation was reported as 55% at centers that performed more than 10 transplants. The 5-year rate was 48% in 1996 and expected to continue to improve over time.^{457a} The 1-year survival rate after liver transplantation for this population is approximately 80%, with beneficial effects on lung function, nutritional status, body composition, and quality of life in most cases.^{92a}

Improved quality of life after lung transplantation remains tightly linked to survival and is difficult to evaluate.

ate. Current information suggests that quality of life increases after lung transplantation for survivors but that it decreases with time and complications such as bronchiolitis obliterans.^{261a}

SPECIAL IMPLICATIONS FOR THE THERAPIST 15-17

Cystic Fibrosis

PREFERRED PRACTICE PATTERNS

4A: Primary Prevention/Risk Reduction for Skeletal Demoralization

4C: Impaired Muscle Performance

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6E: Impaired Ventilation and Respiration/Gas Exchange Associated with Ventilatory Pump Dysfunction or Failure

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Airway Clearance Techniques

The therapist must always be aware that anyone with CF is susceptible to infections, in particular *Burkholderia cepacia*. Care must be taken to avoid transmission via equipment, other patients, or oneself. Handwashing is essential, and high alcohol hand rubs may be more effective.¹⁶³ The therapist will be involved with airway clearance techniques carried out several times per day or as often as the person is able to tolerate it without undue fatigue. Airway clearance techniques should not be performed before or immediately after meals, so treatment must be scheduled to avoid mealtimes.

Aerosol therapy to deliver medication to the lower respiratory tract should be administered just before airway clearance techniques to maximize the effectiveness of both treatments. Breathing exercises, improving posture, mobilizing the thorax through active exercise, and manual therapy are part of promoting good breathing patterns and improving inspiratory muscle endurance. Specifics of airway clearance techniques for this population are beyond the scope of this text; the reader is referred to more detailed materials available.^{102,103,105,140}

The many difficulties surrounding percussion and postural drainage (e.g., poor compliance, time consuming, and requiring the assistance of a trained individual) have resulted in the development of alternative airway clearance techniques that can be accomplished without the assistance of another caregiver.

Each of these techniques (e.g., autogenic drainage, active cycle breathing, positive expiratory pressure [PEP], Flutter valve therapy, Acapella, and Quake) requires a certain level of compliance, motivation, understanding, neuromuscular function, and breath control. The therapist is very instrumental in evaluating each individual's needs, motivation, abilities, resources, and preferences in determining the best intervention or interventions to use.



Figure 15-15

PARI PEP. The positive expiratory pressure (PEP) device maintains pressure in the lungs, keeping the airways open and allowing air to get behind the mucus to improve airway clearance, lung volume capacity, and oxygenation. The device can be used by children (A) and adults (B). (PARI PEP is a registered trademark of PARI GmbH. Used with permission. Please note: the authors have no commercial gain from inclusion of this product.)

Autogenic drainage, active cycle breathing, and PEP help the individual to move the mucus up to the larger airways where it can be coughed out more easily. Autogenic drainage comprises a series of sequential breathing exercises designed to clear the small, medium, and large airways in that order. The PEP device maintains pressure in the lungs, keeping the airways open and allowing air to get behind the mucus (Fig. 15-15). This device has been shown to be effective in increased sputum production, improved lung function, and improved oxygenation.¹⁰⁷

The Flutter valve is similar to the PEP device but utilizes a stainless steel ball that vibrates, alternately opening and closing the device's air hole, pulsing air

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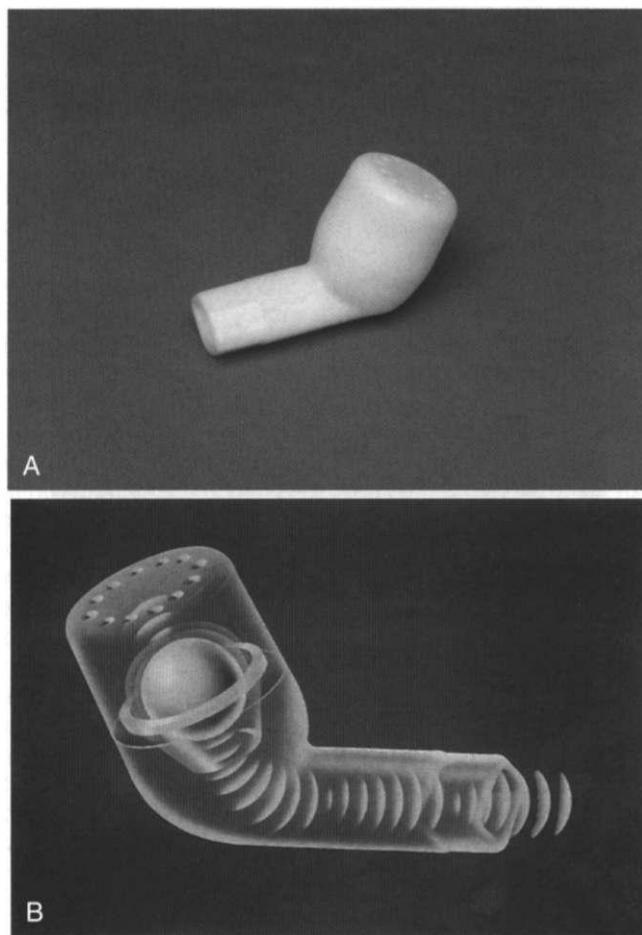


Figure 15-16

The Flutter mucus clearance device. **A**, Flutter device can be used in anyone with chronic obstructive pulmonary disease (COPD). **B**, Flutter valve shown in this schematic utilizes a stainless steel ball that vibrates, alternately opening and closing the device's air hole, pulsing air back into the airways. (Courtesy Axcan Pharma, Birmingham, AL, 2007. Used with permission.)

back into the airways (Fig. 15-16). Current practice is to use one of the devices (PEP or Flutter valve) followed by autogenic breathing techniques. The total treatment time is about 15 minutes and can be carried out independently by some children and most adolescents and adults with mild-to-moderate disease who can follow the directions and control their breathing.

Preliminary studies on the Flutter device suggest that Flutter valve therapy is an acceptable alternative postural drainage and percussion and may be more effective than postural drainage in prolonging the ability to raise secretions. Sputum production increased significantly 30 minutes after the end of treatment, and 1 hour after the end of treatment, it was significantly greater than the amount produced after postural drainage.^{26,170} In a comparison study lasting 1 year, PEP was superior to the Flutter in maintaining pulmonary function, reducing hospitalizations, and reducing antibiotic use.²⁹¹

A high-frequency chest wall oscillation vest can provide mucus clearance for those individuals who

lack the ability to perform the simpler techniques and is especially helpful with children (although the cost may seem prohibitive, it is less than a single hospitalization for a pulmonary exacerbation). The device consists of an inflatable vest (The Vest) attached to an air pulse generator (Fig. 15-17). The generator, a compressor-like device, rapidly inflates and deflates the vest, gently compressing and releasing the chest wall to create airflow within the lungs. This device treats all lobes simultaneously for the duration of the time it is activated. This process moves mucus toward the larger airways where it can be cleared by coughing.

The effectiveness of this mechanized device has been supported by the limited research published to date. Results from The Vest are becoming quite well documented, demonstrating decreased hospitalizations and slower rate of FEV₁ decline.^{255,377} The Vest Clearance System web site (<http://www.thestvest.com/research/bibliography.asp>) has an extensive bibliography. Future research should determine optimal compression frequencies and wave forms.^{255,299,377} The Vest is comparable to PEP in effectiveness, but arterial oxygen saturation may drop during its use, so this must be monitored during exacerbations.¹⁰⁷

Nutrition

Malnutrition and deterioration of lung function are closely interrelated and interdependent in the person with CF. Each affects the other, leading to a spiral decline in both. The occurrence of malnutrition during childhood seems to be associated with impaired growth and repair of the airway walls. In children, when growth in body length occurs, good nutrition is associated with better lung function. When adequate nutrition is combined with physical training and aerobic exercise, improved body weight, respiratory muscle function, lung function, and exercise tolerance occur with increases in both respiratory and other muscle mass.¹⁹²

Exercise

Increasingly, exercise and sport are being advanced as core components of treatment for individuals with CF of all ages. A large portfolio of exercise literature has already established that supervised exercise programs enhance fitness (and thereby improve survival), increase sputum clearance, delay the onset of dyspnea, delay declines in pulmonary function, prevent decrease in bone density,⁴⁴ enhance cellular immune response,⁴⁶ and increase feelings of well-being, thereby potentially improving self-image, self-confidence, and quality of life for the person with CF. Both short- and long-term aerobic and anaerobic training have positive effects on exercise capacity, strength, and lung function.⁵⁵

Reduced systemic oxygen extraction is an important factor, limiting exercise in many individuals with CF, but the specific parameters of this limitation remain unknown and probably vary from person to person.^{312,325} Even unsupervised programs produce a training effect and pulmonary benefits.³⁰⁶ Inspiratory muscle training alone in individuals with CF has shown improved lung function and increased

**Figure 15-17**

A, 4-year-old wearing American Biosystems, Inc (ABI) Vest to self-administer high-velocity oscillations. The vest can accommodate a child as young as 2 years old and is worn 30 minutes twice each day. Medications used to open airways and relax bronchospasms are administered through a nebulizer during the first 10 minutes of treatment, and the machine automatically shuts off every 10 minutes to allow the person to clear secretions. **B**, A foot-pad control mechanism (not shown) can be used to manually stop the machine anytime to allow the client to cough and clear mucus. Any position can be assumed, and at this age, the child can do everything himself. This device promotes compliance and is accompanied by reduced use of medications and infections. (Courtesy Kerry Resch, Missoula, MT, 2001.)

work capacity, as well as improved psychosocial status.^{113,127}

The therapist can be very instrumental in providing client and family education about the importance of combining good nutrition and exercise/activity. The therapist helps each individual develop an exercise routine that includes strengthening, stretching, aerobic, and endurance components with special attention to breathing exercises to aerate all areas of the lungs. Weight loss with exercise is of special concern in this population, especially for the individual with CF and diabetes mellitus.

Energy expenditure is higher than usual for individuals with CF and diabetes during periods of recovery from mild exercise or activity because of increased work of breathing (WOB) consistent with higher ventilatory requirements.⁴⁴¹ This requires careful collaboration among client/family, therapist, and nutritionist. In addition, systemic inflammatory response to exercise may be greater for individuals with CF, potentially exacerbating the disease.²¹³

As longevity increases with CF, quality of life issues are more important, thus these issues, such as posture, arthropathies, and neuromuscular control, are becoming more important for the physical therapist.^{121,285} When treating anyone with CF who has sustained trauma, a multisystem approach is critical for optimal outcomes in physical therapy.⁴¹¹

Individuals with CF awaiting a transplant must remain as active as possible; whenever possible, the therapist can design a safe but effective exercise program. If significant oxygen desaturation or severe breathlessness limits activity, then exercise on a tread-

mill, stationary bike, or even a stationary device for seated pedaling is recommended with supplemental oxygen supplied at sufficient flow to match minute ventilatory requirements.⁴⁴⁴

Studies of exercise performance in lung transplant recipients with end-stage CF report that exercise performance improves after transplantation but remains well below normal.³²⁵ In a study of 12 individuals 8 to 95 months after lung transplant, the diaphragm and abdominal muscle strength was preserved relative to healthy controls, but quadriceps strength was significantly diminished and affected exercise performance. Corticosteroid use partly contributed to this strength deficit.³⁴²

Athletes with Cystic Fibrosis

For those individuals who are able and interested in participating in sports (Fig. 15-18), special considerations must be addressed. Calorie intake and maintaining weight, nutrition, and fluid and electrolyte balance are major concerns. Each individual must be assessed, evaluated, and monitored carefully. The therapist, family, and nutritionist can work together to minimize exercise and nutrition-induced complications. The information presented here is only a general guideline and may need to be modified for individual needs, metabolism, personal health, level and type of sports participation, and so on.

During the off-training season, the individual (especially children and adolescents) will need to eat one and one-half times the protein and calories of an athlete who does not have CF in order to maintain

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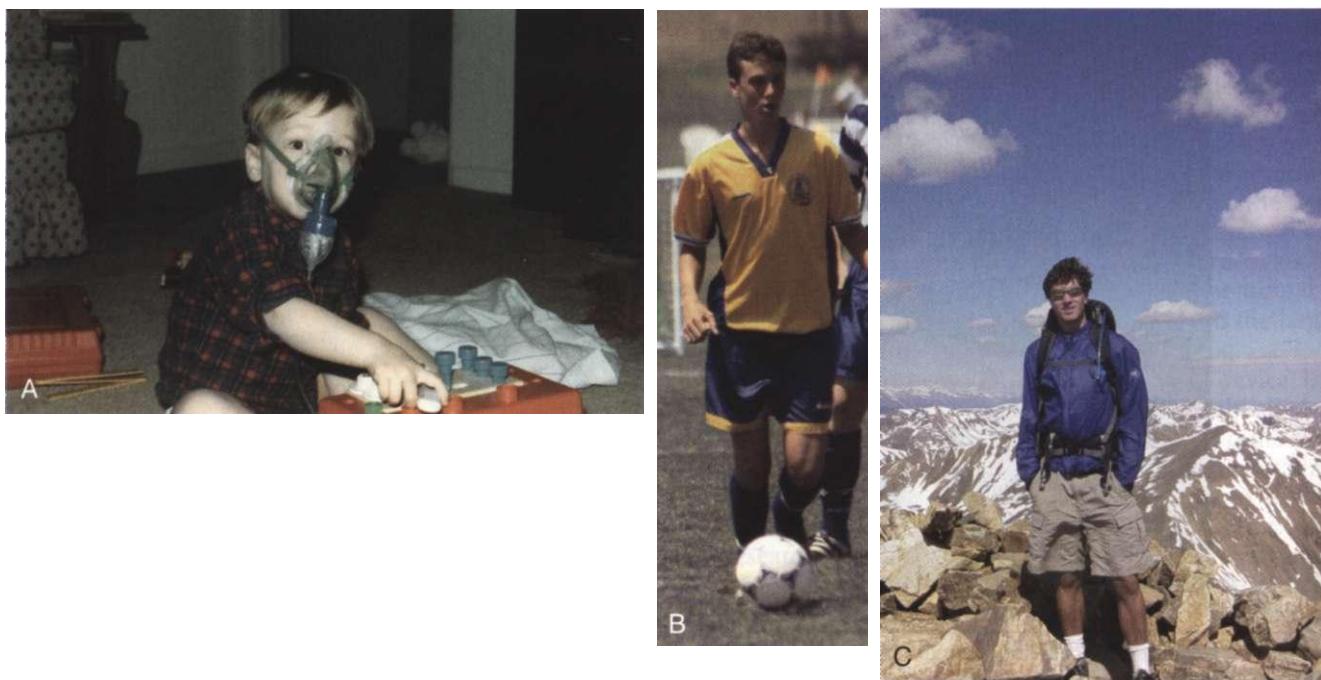


Figure 15-18

A, 18-month-old shortly after diagnosis; face-mask is a nebulizer (device designed to create and throw an aerosol) that is delivering albuterol (bronchodilator). **B**, This same individual in 1999 at age 15 years (6 feet tall; 145 lb) competing in a regional soccer tournament. **C**, Same young man at 23 years (6 feet 2 inches, 175 lb), still actively hiking, cycling, running, and playing intramural sports, while enrolled in graduate school. (Courtesy Kevin Hanson, Helena, MT, 2007. Used with permission.)

weight. During the sport season, calorie intake must be increased with the goal of maintaining weight.

Hyponatremia (deficiency of sodium in the blood; see discussion in Chapter 5) can be a serious problem for athletes with CF who excrete three to five times the sodium (in sweat) of an athlete without CF during sports participation. This situation combined with increased intake of water further dilutes the sodium levels in the body. Sodium loss combined with losses of potassium and magnesium can result in life-threatening situations for these individuals.

Some guidelines for these athletes include weighing before and after exercise and considering the loss as a loss of fluids accompanied by electrolytes; replacing fluid loss with electrolytes (e.g., drinks such as Gatorade or Recharge) instead of water; taking an appropriate number of salt tablets; and eating a meal with sodium-, potassium-, and magnesium-rich foods.

Sporting activities should not be undertaken during infective exacerbations. Sports that carry a medical risk for people with CF (e.g., bungee and parachute jumping, skiing, or scuba diving) should be avoided. Individuals with CF who have portal hypertension with significant enlargement of the spleen and liver should be advised against contact sports, such as rugby and football, in addition to bungee and parachute jumping.

Skiing for anyone with CF who is already hypoxic is not advised; episodes of acute right-sided heart failure brought on by a combination of altitude and

unaccustomed strenuous anaerobic and aerobic exercise have been reported. Scuba diving is contraindicated for anyone with lung disease if there is any evidence of air trapping. On ascent, the air expands, increasing the risk of developing a pneumothorax.⁴⁴⁴

Transition to Adult Care

CF centers and other centers providing life-long care for clients with CF have realized the need for a transition phase between pediatric and adult care and the provision of counseling for parents and the young adult with CF to assist them with this marked change in approach to the young adult's care.

The physical therapist can and should have an integral role in preparing families and clients for various phases in care from pediatrics to adolescent care to a more independent model of adult care. If such a program does not exist in your current facility, materials and resources are available to help get such a program started and established.^{40,85,289,455}

Many families receive care for their child for years at a pediatric center and develop life-long relationships with health care agencies and staff. Every effort should be made to accomplish a smooth transition for the client and the family, since the move from the well-established pediatric facility to a new facility or medical team can be a stressful transition for all.

Families in rural settings or who travel distances to benefit from centralized services in CF clinics or centers have some unique needs that should be addressed as well. For example, maintaining a complete medical

record in more than one facility is not always possible. Without a good systems coordinator, gaps in the medical record from one clinic to another become all too common.

Transition to adult care should be a planned process over time. Every effort should be made to avoid an abrupt transfer. The pediatric team has the responsibility to "set the stage" for the transition. The process can be started early with expectations for an eventual adult transition reinforced in intervals. Written information about the transition, setting an actual time frame, and planning each step in the process are important.²⁸⁹

Three guiding principles are suggested, regardless of the format chosen for transition. First, there must be an adult team that is both interested and able to provide care. Second, there needs to be a very well planned, coordinated approach to the transition. Third, there must be excellent communication and interaction between the pediatric and adult teams.²⁸⁹ Transition programs should be flexible enough to meet the needs of a wide range of young people, health conditions, and circumstances. The actual transfer of care should be individualized to meet the specific needs of young people and their families.³⁶² Family involvement, including parents, guardians, and/or partner and the client, is essential to the success of any transition phase. The omission of any key people from the transition team can result in frustration, feelings of abandonment, and miscommunication, which could ultimately lead to compromised care for the individual with CF.

Adults With Cystic Fibrosis

As individuals with CF survive longer into adulthood, the unique needs of this population group are being considered. Health care in the adult setting encourages independence and increased self-reliance.⁴⁵⁹ Achieving an ideal nutritional status is an integral part of management of people with CF, but how these requirements change as the individual ages remains unknown. Emphasis is continually placed on dietary intake and weight; the effects of this on eating behavior and self-perceptions are under investigation.^{1,424,446}

Other concerns include the effects of long-term use of pancreatic enzymes, osteoporosis associated with late-stage CF and its complications of increased fracture rates and severe kyphosis,^{19,20,191} the effects of hormonal changes in relation to the menstrual cycle on lung function,²²⁴ psychosocial-spiritual issues,²⁰⁰ and infertility issues.²⁰⁹

The origin of bone disease in CF is multifactorial and not completely understood. However, glucocorticoid therapy, delayed pubertal maturation, malabsorption of vitamin D, poor nutritional status, inactivity, and hypogonadism are all potential contributing factors.^{105a} In addition, chronic pulmonary inflammation causes increased circulating levels of cytokines that augment bone resorption and suppress bone formation. Decreases in BMD resulting from these factors can lead to osteoporosis, fragility fractures, and possible exclusion from lung transplant candidacy.^{105a}

Although a number of therapeutic options are now available for osteoporosis, it is likely that prevention, prompt recognition, and early treatment will be far more effective in achieving bone health than intervention at later stages of the problem. An in-depth discussion of bone health and disease in CF is available.^{105a}

Stress urinary incontinence has also been shown to affect many girls and women with CF, probably caused by the forceful and prolonged coughing bouts characteristic of the disease.^{94,294} Interventions aimed at improving pelvic floor muscle strength and coordination may be appropriate for these individuals.

PARENCHYMAL DISORDERS

Atelectasis

Definition

Atelectasis is the collapse of normally expanded and aerated lung tissue at any structural level (e.g., lung parenchyma, alveoli, pleura, chest wall, bronchi) involving all or part of the lung. Most cases are categorized as obstructive-absorptive or compressive.

Etiologic Factors and Pathogenesis

The primary cause of atelectasis is obstruction of the bronchus serving the affected area. If a bronchus is obstructed (e.g., by tumors, mucus, or foreign material), atelectasis occurs as air in the alveoli is slowly absorbed into the bloodstream with subsequent collapse of the alveoli. Atelectasis can also develop when there is interference with the natural forces that promote lung expansion (e.g., hypoventilation associated with decreased motion or decreased pulmonary expansion such as occurs with paralysis, pleural disease, diaphragmatic disease, severe scoliosis, or masses in the thorax). Failure to breathe deeply postoperatively (i.e., because of muscular guarding and splinting from pain or discomfort with an upper abdominal, chest, or sternal incision), oversedation, immobility, coma, or neuromuscular disease can also interfere with the natural forces that promote lung expansion, leading to atelectasis.

Insufficient pulmonary surfactant, such as occurs in respiratory distress syndrome, inhalation of anesthesia, high concentrations of oxygen, lung contusion, aspiration of gastric contents or smoke inhalation, or increased elastic recoil as a result of interstitial fibrosis (e.g., silicosis, asbestosis, radiation pneumonitis), can also interfere with lung distention. When atelectasis is caused by inhalation of concentrated oxygen or anesthetic agents, quick absorption of these gases into the bloodstream can lead to collapse of alveoli in dependent portions of the lung.

Although atelectasis is usually caused by bronchial obstruction, direct compression can also cause it. The compressive type is due to air (pneumothorax), blood (hemothorax), or fluid (hydrothorax) filling the pleural space. Abdominal distention that presses on a portion of the lung can also collapse alveoli, causing atelectasis. *Right middle lobe syndrome* refers to atelectasis secondary

to compression of the bronchus to the right middle lobe by lymph nodes containing metastatic cancer.

Clinical Manifestations

When sudden obstruction of the bronchus occurs, there may be dyspnea, tachypnea, cyanosis, elevation of temperature, drop in blood pressure, substernal retractions, or shock. In the chronic form of atelectasis, the client may be asymptomatic with gradual onset of dyspnea and weakness.

MEDICAL MANAGEMENT

DIAGNOSIS. Atelectasis is suspected in penetrating or other chest injuries. X-ray examination may show a shadow in the area of collapse. If an entire lobe is collapsed, the radiograph will show the trachea, heart, and mediastinum deviated toward the collapsed area, with the diaphragm elevated on that side (see Figs. 15-13 and 15-22). Chest auscultation and physical assessment add to the clinical diagnostic picture. Blood gas measurements may show decreased oxygen saturation.

TREATMENT AND PROGNOSIS. Once atelectasis occurs, treatment is directed toward removing the cause whenever possible. Suctioning or bronchoscopy may be employed to remove airway obstruction. Airway clearance techniques are helpful to remove secretions and promote segmental inflation after the obstruction has been removed.

Surfactant has been used to resolve atelectasis in infants with respiratory distress syndrome, meconium aspiration, and other pathologies.¹³² Antibiotics are used to combat infection accompanying secondary atelectasis. Reexpansion of the lung is often possible, but the final prognosis depends on the underlying disease. Chronic atelectasis may require surgical removal of the affected segment or lobe of lung.

SPECIAL IMPLICATIONS FOR THE THERAPIST

Atelectasis

PREFERRED PRACTICE PATTERNS

See also Special Implications for the Therapist: Pneumothorax and Special Implications for the Therapist: Chest Wall Disease or Injury in this chapter.

6B: Impaired Aerobic Capacity/Endurance Associated with Deconditioning

6C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated with Airway Clearance Dysfunction

6F: Impaired Ventilation and Respiration/Gas Exchange Associated with Respiratory Failure

Atelectasis is a postoperative complication of thoracic or high abdominal surgery. Left lower lobe atelectasis can occur following cardiac surgery. Within a few hours after surgery, atelectasis becomes increasingly resistant to reinflation. This complication is exacerbated in people receiving narcotics. Although there has not been sufficient research to support its use, one

of the goals of acute care therapy is to prevent atelectasis in the high-risk client.³³⁵

Diminished respiratory movement as a result of postoperative pain is often addressed by the therapist. Frequent, gentle position changes, deep breathing, coughing, and early ambulation help promote drainage of all lung segments. Deep breathing and effective coughing enhance lung expansion and prevent airway obstruction. Deep breathing is beneficial because it promotes the ciliary clearance of secretions, stabilizes the alveoli by redistributing surfactant, and permits collateral ventilation of the alveoli, through Kohn's pores in the alveolar septa.

Kohn's pores, which open only during deep breathing, allow air to pass from well-ventilated alveoli to obstructed alveoli, minimizing their tendency to collapse and facilitating removal of the obstruction. To minimize postoperative pain during deep-breathing and coughing exercises, teach the client to hold a pillow firmly over the incision.

There is evidence that vigorous airway clearance techniques are as effective in resolving atelectasis from mucous plugs as bronchoscopy.²⁵⁰ Bronchoscopy has the adverse effect of temporarily lowering oxygen saturation and further irritation of lung tissue.

Pulmonary Edema

Definition and Incidence

Pulmonary edema or pulmonary congestion is an excessive fluid in the lungs that may accumulate in the interstitial tissue, in the air spaces (alveoli), or in both. The fluid is a barrier to gas exchange. Pulmonary edema is a common complication of many disease processes. It occurs at any age but with increasing incidence in older people with left-sided heart failure.

Etiologic and Risk Factors

Most cases of pulmonary edema are caused by left ventricular failure (see Fig. 12-12, A), acute hypertension, or mitral valve disease, but noncardiac conditions, especially kidney or liver disorders prone to the development of sodium and water retention, can also produce pulmonary edema. These causes of pulmonary edema include intravenous narcotics, increased intracerebral pressure, brain injury, high altitude, diving and submersion, sepsis, medications, inhalation of smoke or toxins (e.g., ammonia), blood transfusion reactions, shock, and disseminated intravascular coagulation.^{32,90,146}

Other risk factors include hyperaldosteronism, Cushing's syndrome, use of glucocorticoids, and use of hypotonic fluids to irrigate nasogastric tubes. Pulmonary edema itself is a major predisposing factor in the development of pneumonia that complicates heart failure and ARDS.

Pathogenesis

Pulmonary edema occurs when the pulmonary vasculature fills with fluid that leaks into the alveolar spaces,