MMWR™

Morbidity and Mortality Weekly Report

Surveillance Summaries November 11, 2005 / Vol. 54 / No. SS-5

Pulmonary Hypertension Surveillance — United States, 1980–2002

DEPARTMENT OF HEALTH AND HUMAN SERVICES
CENTERS FOR DISEASE CONTROL AND PREVENTION

The *MMWR* series of publications is published by the Coordinating Center for Health Information and Service, Centers for Disease Control and Prevention (CDC), U.S. Department of Health and Human Services, Atlanta, GA 30333.

SUGGESTED CITATION

General: Centers for Disease Control and Prevention. Pulmonary Hypertension Surveillance — United States, 1980–2002. In: Surveillance Summaries, November 11, 2005. MMWR 2005;54(No. SS-5).

Specific: [Author(s)]. [Title of particular article]. In: Surveillance Summaries, November 11, 2005. MMWR 2005;54(No. SS-5):[inclusive page numbers].

CONTENTS

Pulmonary Hypertension Surveillance — United States, 1980–2002

Alexandra Hyduk, MPH
Janet B. Croft, PhD
Carma Ayala, PhD
Kan Zheng
Zhi-Jie Zheng, MD, PhD
George A. Mensah, MD
Division for Heart Disease and Stroke Prevention
National Center for Chronic Disease Prevention and Health Promotion

Abstract

Problem/Condition: Pulmonary hypertension, which is listed on hospital records and death certificates as either primary (i.e., idiopathic) pulmonary hypertension or pulmonary hypertension secondary to another underlying condition or disease, is considered rare amongst the U.S. population. Limited reports have been published regarding surveillance data for this debilitating and often fatal condition.

Reporting Period Covered: 1980–2002.

Description of Systems: This report summarizes mortality data from the National Vital Statistic System (NVSS) and hospital discharge data from the National Hospital Discharge Survey (NHDS) for 1980–2002 and Medicare hospital claims data for 1990–2002. NVSS, maintained by CDC's National Center for Health Statistics (NCHS), compiles multiple cause of death information from official death certificates filed in the United States. NHDS, also conducted annually by NCHS, includes information on discharges from a sample of nonfederal, short-stay hospitals. Annual hospital claims and enrollment data for all Medicare beneficiaries aged ≥65 years are provided by the Center for Medicare and Medicaid Services. Because pulmonary hypertension might be reported secondary to other diseases, this report presents data for pulmonary hypertension as any contributing cause of death or any-listed hospital diagnosis.

Since 1980, the numbers of deaths and hospitalizations, death rates, and hospitalization rates have increased for pulmonary hypertension, particularly among women and older adults. During 1980–2000, death rates were higher for men than women; however, by 2002, no difference in rate was observed because of increasing death rates among women and declining death rates among men. Hospitalization rates were higher for men than for women until 1995; after 1995, higher rates were observed among women. Death rates since 1985 and Medicare hospitalization rates throughout the reporting period 1990–2002 have been higher for blacks than for whites. In addition, two distinct geographic clusters were observed for the highest hospitalization rates in the Medicare population and the highest death rates for pulmonary hypertension, in the western United States and in the Appalachian region.

Interpretation: Increases in mortality from and hospitalization for pulmonary hypertension might reflect increased physician awareness and changes in diagnosing and reporting this chronic disease. Although pulmonary hypertension historically has been considered a disease of women of childbearing age, it affects all ages and racial populations. Older women represent the majority of patients and decedents with this condition. More research is needed concerning cause, prevention, and treatment of pulmonary hypertension. Public health initiatives should include increasing physician awareness that early detection is needed to initiate prompt, effective disease management. Additional epidemiologic initiatives also are needed to ascertain prevalence and incidence of various pulmonary hypertension disease entities such as pulmonary arterial hypertension.

This material in this report originated in the National Center for Chronic Disease Prevention and Health Promotion, Janet L. Collins, PhD, Director; and the Division for Heart Disease and Stroke Prevention, Darwin R. Labarthe, MD, Acting Director.
Corresponding author: Janet B. Croft, PhD, National Center for Chronic Disease Prevention and Health Promotion, 4770 Buford Hwy, NE, Atlanta, GA 30341, MS K-47. Telephone: 770-488-2424; Fax: 770-488-8151; E-mail: JCroft@cdc.gov.

Introduction

Pulmonary hypertension is a hemodynamic state signifying the presence of one or more pulmonary vascular disease entities and is characterized by a severe elevation in the mean pulmonary arterial pressure and pulmonary vascular resistance (*1*). Because early symptoms (i.e., difficulty in breathing and fatigue) and later manifestations (i.e., reduced exercise tolerance, palpitations, fainting, swelling of the ankles or legs, chest pain, or hoarseness) are symptomatic of other diseases, prompt diagnosis and treatment can be delayed because physicians often have to exclude other causes for these symptoms (*2–4*). Pulmonary hypertension is a devastating chronic disease with a poor long-term prognosis; results from a national registry of patients with primary (i.e., idiopathic) pulmonary hypertension indicated that the duration from onset of symptoms to death was an average of 2.8 years (*5*). In recent years, new treatments have become available for primary pulmonary hypertension, particularly for pulmonary arterial hypertension, which include anticoagulants, calcium channel blockers, and prostacyclins (*3,4,6–12*); these treatments prolong survival and provide clinical improvement but can often be expensive. Although effective management might be possible with early detection, the majority of persons have pulmonary hypertension diagnosed in later stages of the disease, making treatment more difficult and less successful.

Pulmonary hypertension is reported on hospital records and death certificates as either primary pulmonary hypertension or as pulmonary hypertension secondary to another underlying condition or disease. Pulmonary hypertension might be secondary to congenital heart disease, valvular heart disease, chronic thromboembolic disease, lung diseases, liver diseases, sleep-disordered breathing and hypoxemia, lupus, scleroderma, rheumatoid arthritis, vasculitis, or human immunodeficiency virus (HIV) infection (*3,7,13*). Traditionally, primary pulmonary hypertension might have been diagnosed when no obvious cause of death could be identified. The traditional classification of primary or secondary pulmonary hypertension has limitations and has not been useful in clinical practice (*4*). In 1998, a conference sponsored by the World Health Organization proposed a new treatment-oriented classification of all pulmonary hypertensive diseases rather than addressing only primary pulmonary hypertension (*4,14*). Therefore, this report will focus on all forms of pulmonary hypertension.

Limited national statistics are available regarding pulmonary hypertensive diseases (*15,16*). Although a rare condition, death rates for primary pulmonary hypertension as the underlying cause of death have increased since 1979 (*15*), and the number of all cases is likely higher than that reported because of difficulties in disease detection (*16*). This report describes trends during 1980–2002 in diagnosed pulmonary hypertension–related deaths and hospitalizations, the only national surveillance data available for pulmonary hypertension. Because pulmonary hypertension might be more likely to be reported secondary to other diseases, this report presents data for pulmonary hypertension as any contributing cause of death or as any-listed hospital diagnosis.

Methods

For this analysis, pulmonary hypertension included primary pulmonary hypertension (i.e., idiopathic pulmonary arteriosclerosis, idiopathic or primary pulmonary artery hypertension, and essential or idiopathic pulmonary hypertension); secondary pulmonary hypertension; other specified pulmonary heart disease, including a small number of cardiopulmonary events; and unspecified chronic pulmonary heart disease, including chronic cardiopulmonary disease and chronic cor pulmonale (*17–19*). To examine trends in pulmonary hypertension, CDC analyzed data from the National Vital Statistics System (NVSS), the National Hospital Discharge Survey (NHDS), and Medicare hospital claims and enrollment records.

Mortality Data

NVSS, which is maintained by the National Center for Health Statistics (NCHS), compiles data from vital records on all deaths occurring annually in the United States. Public use data from NVSS files provide the underlying cause of death and up to 19 additional contributing causes and demographic data on decedents (*20*). Demographic data on death certificates are reported by funeral directors, usually from information provided by family members of decedents. A physician, medical examiner, or coroner reports on the death certificate the diseases, injuries, or complications that caused the death. The underlying cause of death is the disease (or injury) that initiated the sequence of events leading directly to death. For this analysis, all diseases and conditions reported on death certificates were classified according to codes from the *International Classification of Diseases, Ninth Revision* (ICD-9) for deaths during 1980–1998 (*17*) and codes from the *International Classification of Diseases, Tenth Revision* (ICD-10) for deaths during 1999–2002 (*18,19*). Pulmonary hypertension was diagnosed for decedents with ICD-9 codes 416.0, 416.8, or 416.9 during 1980–1998 or ICD-10 codes I27.0, I27.8, or I27.9 during 1999–2002 reported as any contributing cause of death (i.e., any of the possible 20 conditions, including underlying cause) on the death certificate.

Numbers of U.S. residents for the period 1980–2002 were provided by the U.S. Bureau of the Census and used to calculate death rates per 100,000 population. Bridged-race Census estimates were used as denominators for groups defined by

race and Hispanic origin during 1990–2002 (*20*). Death rates were age-standardized to the 2000 U.S. standard population (*21,22*). Age-specific death rates were calculated for groups aged <45, 45–54, 55–64, 65–74, 75–84, and ≥85 years. Before 1990, race categories on Census data included only white, black, and other race; beginning in 1990, race categories included white, black, Asian/Pacific Islander, and American Indian/Alaska Native. Hispanic origin was also included in Census data beginning in 1990. Because numbers of deaths with pulmonary hypertension as a contributing cause were relatively small each year, data were aggregated into four 5-year periods (1980–1984, 1985–1989, 1990–1994, and 1995–1999) and one 3-year period (2000–2002) to examine decedent characteristics and age-specific and -standardized death rates among groups defined by race, sex, or age. For 2000–2002, data were aggregated for all 50 states and the District of Columbia (DC) to examine geographic variations in deaths with pulmonary hypertension as a contributing cause.

The distribution of selected disease categories reported as the underlying cause of death among decedents with reported pulmonary hypertension was examined for each period. These disease categories were selected based on reported associations of particular diseases with pulmonary hypertension. A major category of interest was chronic lower respiratory disease (e.g., chronic bronchitis, unspecified bronchitis, emphysema, asthma, and other chronic lower respiratory diseases), of which encompasses chronic obstructive pulmonary disease (*23*).

Hospitalizations

NHDS is conducted annually by NCHS and provides demographic and discharge information abstracted from medical records of inpatients selected from a nationally representative sample of nonfederal, short-stay hospitals in the 50 states and DC (*24,25*). Only hospitals with six or more staffed beds and an average length of patient stay of <30 days or those with a general (i.e., medical or surgical) specialty or children's specialty were included in the survey (*25*). Estimates of the number and rate of hospitalizations attributed to pulmonary hypertension (*International Classification of Diseases, Ninth Revision, Clinical Modification* [ICD-9-CM] codes 416.0, 416.8, or 416.9 [*26*]) are provided in this report. These estimates are based on counting a patient if a diagnosis is determined as any one of the up to seven medical diagnoses recorded in NHDS. Estimates from NHDS were calculated by using the weights (i.e., inflation factors) that allow estimation from the sample.

Data on the U.S. civilian population for the period 1980–2002 were obtained from the U.S. Bureau of the Census and used to calculate age- and sex-specific hospitalization rates per 100,000 population. Age-specific hospitalization rates were calculated for persons aged <45, 45–54, 55–64, 65–74, 75–84, and ≥85 years. Hospitalization rates for pulmonary hypertension as any-listed diagnosis were age-standardized to the 2000 U.S. standard population (*21,22*). To examine trends in hospitalizations during 1980–2002, data were aggregated into four 5-year periods (1980–1984, 1985–1989, 1990–1994, and 1995–1999) and one 3-year period (2000–2002). Race-specific estimates are not provided because of incomplete reporting of race on hospital records (*27*). In addition, the sampling design for NHDS does not provide for state-specific estimates of hospitalizations. The distribution of selected disease categories as the principal (i.e., first listed) diagnosis also was examined for each period among hospitalizations with any-listed pulmonary hypertension.

Medicare Claims

Medicare hospital claims represent the only available source of hospital discharge information for racial populations and all states and DC. All Medicare hospital claims and enrollment record data for 1990–2002 were obtained from the Centers for Medicare and Medicaid Services. A hospitalization for pulmonary hypertension was defined as one for which any of up to 10 diagnoses (five diagnoses in 1990) on the hospital claim was classified as an ICD-9-CM code 416.0, 416.8, or 416.9. The number of persons at risk (i.e., U.S. residents in the 50 states and DC aged ≥65 years who were entitled to Medicare Part A benefits on July 1 during 1990–2002, excluding members of health maintenance organizations) was obtained from Medicare enrollment records. Hospitalization rates per 100,000 Medicare enrollees were age-standardized to the 2000 U.S. standard population aged ≥65 years (*21,22*). Age-specific hospitalization rates were calculated for groups aged 65–74, 75–84, and ≥85 years. Race categories on hospital claims and enrollment records included white, black, Asian, Hispanic, and American Indian. State-specific numbers were obtained, and age-standardized rates of hospitalizations with pulmonary hypertension as any-listed diagnosis were calculated for the Medicare population in all 50 states and DC.

Results

Mortality Data

During 1980–2002, the total number of deaths attributed to pulmonary hypertension as any contributing cause of death increased from 10,922 to 15,668 (Table 1); however, the increase was observed only among women. During 1980–1991, women had lower numbers of deaths from pulmonary hypertension than men; after 1991, women had higher numbers of

deaths than men. Age-standardized death rates for the total U.S. population remained relatively stable from 1980 (5.2 per 100,000) through 2002 (5.4 per 100,000) (Table 1). However, rates increased among women but decreased among men during this period such that the age-standardized death rates were similar beginning in 2000 (Figure 1). Among all racial populations, the number of deaths from pulmonary hypertension reported as any contributing cause of death increased, with blacks having the highest age-standardized death rates (Table 2, Figure 2). Non-Hispanics had higher age-standardized death rates than Hispanics (Table 3). Age-specific death rates for pulmonary hypertension reported as any contributing cause of death also increased, primarily among adults aged ≥85 years, particularly during 1990–2002 (Table 4, Figure 3).

The annual number of deaths from pulmonary hypertension as any contributing cause of death was aggregated into the five selected periods (1980–1984, 1985–1989, 1990–1994, 1995–1999, and 2000–2002) for groups defined by sex, age, race, and Hispanic ethnicity to obtain stable estimates for comparisons. Over the five periods, age-standardized death rates for pulmonary hypertension as any contributing cause of death increased among women and blacks (Table 5). Age-specific death rates increased among men aged ≥85 years, women aged ≥65 years, whites aged ≥75 years, and blacks aged ≥65 years (Table 5). Nevertheless, during 2000–2002, men had slightly higher age-specific death rates than women in the older age groups (Figure 4). At ages <75 years, blacks had higher age-specific death rates than whites during 2000–2002, but whites had higher death rates than blacks at ages ≥85 years (Figure 5).

During 2000–2002, age-standardized death rates for pulmonary hypertension reported as any contributing cause of death varied among states/areas (Table 6). States/areas with the highest age-standardized death rates were Wyoming (9.9), Idaho (8.9), DC (8.8), Colorado (8.6), Vermont (8.4), Alaska (7.7), Montana (7.4), North Carolina (7.4), Ohio (7.4), West Virginia (7.3), and New Hampshire (7.2).

During 1980–2002, because of increased numbers of death among women, the proportion of decedents with reported pulmonary hypertension who were female increased. During 2000–2002, 58.3% of all decedents with pulmonary hypertension reported on their death certificates were female (Table 7). Among decedents with reported pulmonary hypertension, the proportion whose race was reported as black also increased.

Among decedents with pulmonary hypertension reported, little change was observed in the proportions of those who were aged <55 years. However, the proportion of decedents aged ≥75 years increased during 1980–2002 such that 30.6% of all decedents with pulmonary hypertension reported during 2000–2002 were aged 75–84 years and 18.1% were aged ≥85 years, compared with 23.7% and 6.5%, respectively, during 1980–1984 (Table 7). Age distributions were similar among females and males aged <55 years during 2000–2002 (Figure 6); however, 21% of all female decedents with pulmonary hypertension reported were aged ≥85 years, compared with 14% of male decedents, although greater proportions of men than women were aged 65–74 years. Only 11.1% of male decedents and 9.6% of female decedents with pulmonary hypertension reported during 2000–2002 were aged <45 years, an age category that includes women of childbearing age affected by pregnancy-related pulmonary hypertension and newborns and young children with congenital anomalies or familial pulmonary hypertension. In contrast, a higher proportion of decedents were aged <45 years among blacks compared with whites (22.4% versus 8.0%) during 2000–2002 (Figure 7). The proportions who were aged 45–54 years and 55–64 years were higher among blacks than among whites; in contrast, the proportion who were aged ≥65 years was higher among whites than among blacks during 2000–2002 (75.1% versus 48.2%, respectively).

Some disease categories that are commonly reported with pulmonary hypertension were rarely reported as the underlying cause of death among decedents (Table 8). During 2000–2002, among decedents with pulmonary hypertension reported on the death certificate, the most commonly reported underlying cause was pulmonary hypertension (30.3%), followed by chronic lower respiratory disease (25.9%). During 1980–2002, the proportions of decedents with pulmonary hypertension reported as the underlying cause of death increased among all decedents with pulmonary hypertension reported as any cause on the death certificate. Although chronic lower respiratory disease was the most commonly reported underlying cause of death for persons with pulmonary hypertension during 1980–1984 (49.2%), the proportion decreased during 1980–2002. The increase in reported pulmonary hypertension and the decline in reported chronic lower respiratory disease as the underlying cause of death among all decedents with pulmonary hypertension reported were observed in all groups defined by sex, race, and age. Among decedents aged <45 years, the most common underlying causes of death were pulmonary hypertension; congenital malformations; complications of pregnancy, childbirth, and the puerperium, or conditions originating in the perinatal period; and other cardiovascular diseases (Table 9).

Hospitalizations

During 1980–2002, the estimated number of hospitalizations with pulmonary hypertension as any-listed diagnosis tripled for the total U.S. population (Table 10). Compared with estimated numbers of hospitalizations in 1980, the numbers in 2002 were two times higher among men and four times higher among women. Age-standardized hospitalization rates doubled for men and tripled for women. Rates of hospitalizations for pulmonary hypertension among women were higher than those for men, only after 1995 (Figure 8). In 2002, the age-standardized hospitalization rate for pulmonary hypertension as any-listed diagnosis was 95.3 for women and 82.3 for men. Numbers of hospitalizations and age-specific hospitalization rates increased for all age groups in the total U.S. population (Table 11). The greatest increase in hospitalization rate was among adults aged ≥75 years (Figure 9).

To examine trends by sex and age, sample sizes were increased to obtain more reliable estimates by aggregating years of data from NHDS. During 1980–2002, the greatest increase was for ages ≥85 years among men and for ages ≥65 years among women (Table 12). Because of major increases in hospitalizations among women aged ≥85 years, during 2000–2002, women had higher hospitalization rates than men in this older age group (Figure 10).

During 1980–2002, because of substantial increases in the number of women hospitalized with pulmonary hypertension, the proportion increased such that 60.6% of all hospitalizations with any-listed diagnosis of pulmonary hypertension during 2000–2002 were among women (Table 13). During the 2 decades, the proportion of patients aged ≥65 years increased from 55.4% during 1980–1984 to 66.0% during 2000–2002 among all hospitalizations with pulmonary hypertension as any-listed diagnosis. During 2000–2002, the proportion of hospitalized persons aged ≥85 years was higher among women than men (Figure 11).

During 1980–2002, trends in the most commonly reported principal diagnosis changed (Table 14). During 1980–1984, the most commonly reported principal diagnoses among hospitalizations with any-listed diagnosis of pulmonary hypertension were chronic lower respiratory diseases (42.0%), followed by pulmonary hypertension (12.8%) and other cardiovascular diseases (6.8%). By 2000–2002, heart failure (18.7%) was the most commonly reported principal diagnosis, followed by chronic lower respiratory diseases (12.9%) among hospitalizations with any-listed diagnosis of pulmonary hypertension. During 2000–2002, pulmonary hypertension was the principal diagnosis of only 4.2% of hospitalizations involving pulmonary hypertension as any-listed diagnosis. Declines in reporting pulmonary hypertension and chronic lower respiratory disease and the increase in reporting heart failure as the principal diagnosis in these cases were observed among all groups defined by sex and age. Among persons aged <45 years, the principal diagnosis was more likely to be pulmonary hypertension (26.0%) and congenital malformations (22.9%) during 1980–1984; however, by 2000–2002, these conditions had declined as the principal diagnosis among hospitalizations with pulmonary hypertension as any-listed diagnosis (Table 15). Other cardiovascular diseases (29.8%), other respiratory diseases (9.8%), pulmonary hypertension (9.5%), and chronic lower respiratory diseases (7.1%) were the major principal diagnoses during 2000–2002. During this period, other frequently reported principal diagnoses were examined for these pulmonary hypertension hospitalizations among patients aged <45 years. Influenza and pneumonia (4.6%), congenital malformations (4.3%), complications related to specific procedures (3.9%), cellulites and abscess (3.6%), and complications related to pregnancy and childbirth (3.2%) also were principal diagnoses for these younger patients.

Medicare Claims

During 1990–2002, the annual number of hospitalizations for pulmonary hypertension as any-listed diagnosis tripled among Medicare enrollees aged ≥65 years, from 55,516 to 187,205 (Table 16). The age-standardized hospitalization rate was 197.8 in 1990 and 649.7 in 2002. Men had higher age-standardized Medicare hospitalization rates with any-listed pulmonary hypertension than women until 1999 (Figure 12). Increases in numbers of Medicare hospitalizations for pulmonary hypertension were observed among all groups defined by race (Table 17) and age (Table 18). Age-standardized hospitalization rates were higher for blacks than whites (Figure 13). Age-specific hospitalization rates increased for all age groups, but the rates were not the highest among adults aged ≥85 years until after 1995 (Figure 14). By 2000–2002, age-specific hospitalization rates were higher among women at ages 65–74 years and 75–84 years, and whites had higher age-specific hospitalization rates than blacks at ages ≥85 years (Table 19).

During 1990–2002, age-standardized Medicare hospitalization rates increased each year in each state. State data were aggregated for 2000–2002 to obtain more stable Medicare hospitalization rates (Table 20). During 2000–2002, states/areas with the highest age-standardized hospitalization rates with pulmonary hypertension as any-listed diagnosis were Wyoming (1172.1), Colorado (1022.8), West Virginia (954.7), Pennsylvania (907.0), Maryland (841.1), Montana (833.2), Idaho (796.3), New Mexico (792.5), Virginia (788.3), DC (782.0), and Delaware (781.7).

Discussion

The findings in this report indicate that, during 1980–2002, death rates and hospitalization rates for pulmonary hypertension as either any contributing cause of death or as any-listed hospital diagnosis increased. The number of pulmonary hypertension–related deaths and number of hospitalizations also increased, particularly among women, blacks, and older adults. Nevertheless, death rates were higher for men than women during the 2 decades, and hospitalization rates were higher for men than women until 1995. A geographic clustering of the highest Medicare hospitalization rates and highest death rates for pulmonary hypertension was observed in the western United States (in Colorado, Idaho, Montana, and Wyoming) and a second clustering in the Appalachian region (in DC, Maryland, Pennsylvania, Virginia, and West Virginia). In addition, reporting of pulmonary hypertension as the underlying cause of death increased, and reporting pulmonary hypertension as the principal diagnosis decreased during 1980–2002. Among all decedents and all persons hospitalized with pulmonary hypertension, the reporting of chronic lower respiratory disease (including chronic obstructive pulmonary disease) as the underlying cause of death or principal hospital diagnosis declined, and reporting heart failure as the principal diagnosis on the hospital record increased for all age groups.

Increases in reporting pulmonary hypertension as any-listed diagnosis on hospital records might indicate an actual increase in the number of patients or, more likely, a greater increased awareness among physicians to evaluate and diagnose pulmonary hypertension. In 1996, studies suggested a link between appetite suppressants and increases in primary pulmonary hypertension (*28–30*), which probably created greater public and professional interest in the disease, particularly in terms of cases among women. Hospitalization rates for pulmonary hypertension began increasing among both men and women during the early 1990s at the same time that use of appetite suppressants was prevalent (*31*); however, the major increases in hospitalization rates with pulmonary hypertension as any-listed diagnosis seem to have occurred among older women than among younger and middle-aged women who were more likely to take suppressants (*32*).

As the reporting of chronic lower respiratory disease as the underlying cause of death and principal hospital diagnosis declined, heart failure as the principal diagnosis on hospital records and the reporting of pulmonary hypertension as the underlying cause of death on death certificates increased. Changes in the reporting of pulmonary hypertension as the underlying cause of death might be related to changes in medical opinion and interpretation of certification instructions. In addition, increased perception of a condition as serious or fatal could lead to increased reporting of that condition as a contributing cause of death (*33*) or hospitalization. Heart failure is rarely reported on death certificates as the underlying cause because specific instructions on the death certificate state that "cardiac failure" should not be listed as the underlying cause of death. Whether these patterns reflect reporting trends on the part of physicians and health-care systems or actual changes in the etiology and prevalence of pulmonary hypertension should be investigated further. Nevertheless, efforts to eliminate underdiagnosis of pulmonary hypertension and improve the reporting on death certificates and hospital records must be systematic to obtain a more credible estimate of the true distribution of pulmonary hypertension in the U.S. population.

At least three possible explanations exist for the geographic clusterings of Medicare hospitalizations and death rates for pulmonary hypertension in the western United States and Appalachian region. First, differences may exist in medical opinion on certification instructions or on the number of causes to be reported per death certificate. Second, these cases might be more likely to be detected by physicians affiliated with recognized pulmonary hypertension centers of excellence and referral that are located in Denver (CO), Baltimore (MD), and Philadelphia (PA). Third, particular genetic factors or environmental toxins might be more prevalent in those regions. For example, higher hospitalization rates for pulmonary hypertension in the Medicare population in states bordering the Appalachian region might reflect higher proportions of the population who were employed in the coal-mining industry decades ago and who might have contracted respiratory diseases such as "coal-workers" pneumoconiosis, diseases that predispose to pulmonary hypertension. In addition, altitude might also play an important role in the development of pulmonary hypertension in states such as Colorado, Montana, and Wyoming. Identification of these unreported clusters warrants further investigations.

The findings in this report are subject to at least six limitations. First, mortality data are subject to diagnosis and reporting errors by physicians, medical examiners, and coroners, particularly in the absence of an autopsy. Second, the current ICD classifications do not allow data users or those reporting the causes of death and diagnoses to differentiate specific diseases that are of interest in the study of pulmonary hypertension. For example, approximately 99% of all heart failure diagnoses documented in this report were congestive heart failure, a category which also includes right heart failure secondary to left heart failure; however, isolated right heart failure and isolated left heart failure do not have reportable codes but could be of greater interest to future investigators. Similarly, the current ICD codes do not allow differentiation of

the five major categories of pulmonary hypertension entities (Table 21). Thus, population estimates for pulmonary arterial hypertension cannot be ascertained, although this category might be of greater interest to current and future research. Third, NHDS does not include patients admitted to government hospitals (i.e., Veterans Affairs or military); therefore, results might underestimate the total number of hospitalizations for pulmonary hypertension among adults. Fourth, the use of hospital discharge records and Medicare claims records includes the possibility that trends in hospitalization rates could reflect patterns in financial incentives to report other diseases as the principal diagnosis rather than pulmonary hypertension. Fifth, hospital discharge data should not be used to represent prevalence or incidence of the disease because distinguishing between first or new hospitalizations or multiple readmissions during the year is not possible. Finally, a limitation of hospital discharge records and Medicare hospital claims is the inability to determine the accuracy of physician or administrative reporting or the validity of ICD classification of the various disease entities; no information on results of clinical diagnostic evaluations is available. Whether pulmonary hypertension is rare is debatable given the likely underdiagnosis and, thus, underreporting of persons with the disease.

Pulmonary hypertension has had an unclear pathogenesis (*13,34–37*). Although advances have occurred in understanding the pathogenesis of pulmonary hypertension among specialists in the past decade, this understanding might not have reached general practice. In 1998, a World Health Organization symposium developed a more inclusive and clinically useful treatment-based classification system that divided the causes of pulmonary hypertension into five distinct categories of disease entities (*5,14*). Unfortunately, this classification system has not been translated into useful ICD classifications for reporting and distinguishing specific disease entities for surveillance purposes. In 2003, another symposium assessed the impact and usefulness of this classification and determined that, although the new classification was accepted and widely used among specialists and in centers with the largest clinical experience, less experienced physicians apparently continued to use the old classification (i.e., primary versus secondary) (*38*). Therefore, this could complicate prompt evaluation and diagnosis and lead to further misdiagnosis and inaccurate reporting on death certificates and hospital records. Continued clinical research is underway to better understand pulmonary hypertension and its risk factors, as well as the prevention and treatment of pulmonary hypertension. However, professional education efforts should be considered to prompt physician recognition of the early symptoms of suspected pulmonary hypertension, to inform physicians about the treatment-based classification of disease entities, and to educate physicians about the multiple diagnostic evaluations necessary for the categorization and appropriate treatment of patients.

A 2-year delay can occur between onset of symptoms and diagnosis (*12*); however, new treatment modalities have been determined to improve the prognosis of this disease (*11*). In addition to the use of anticoagulants and vasodilators, atrial septostomy, and lung transplants as therapeutic approaches for various pulmonary hypertension entities (*8–12,35,39–41*), clinical trials since 2000 have demonstrated therapeutic efficacy with prostaglandins and endothelin antagonists for pulmonary arterial hypertension (*37,42*). Correct diagnosis of the distinct disease entities of pulmonary hypertension provides direction for management of the underlying disease (*39,43*) and is imperative to prevent a treatment regimen that might worsen the condition or create serious sequelae (*3*). Disease assessment includes physical examination, electrocardiogram, echocardiogram, chest radiograph, ventilation-perfusion scintigraphy, and other assessments (*2,4,10,11,43*). Limitations develop as the symptoms become worse, particularly for symptoms of other comorbid conditions. For example, among patients with HIV, symptoms related to pulmonary hypertension might overshadow the overall clinical problems of these patients (*44*). In addition, pulmonary hypertension might increase the risk for complications and death during pregnancy (*45–47*).

Conclusion

The high proportion of pulmonary hypertension–related deaths and hospitalizations that occurred among adults aged ≥65 years suggested that as the proportion of older adults in the U.S. population increases, pulmonary hypertension might continue to be a more frequent diagnosis, particularly with concomitant chronic heart failure. Because the majority of patients with pulmonary hypertension are older adults, the burden of chronic disability and morbidity on the Medicare system and families will increase. Prevention efforts, including broad-based public health efforts to increase awareness of pulmonary hypertension and to foster appropriate diagnostic evaluation and timely treatment from health-care providers, should be considered. The science base for the etiology, pathogenesis, and complications of pulmonary hypertension disease entities must be further investigated to improve prevention, treatment, and case management. Additional epidemiologic initiatives also are needed to ascertain the prevalence and incidence of the various disease entities.

Although multiple predisposing factors and associated conditions have been identified for pulmonary hypertension, the causal roles and strengths of association have not been well established (*38,48*). Therefore, it is not possible to establish preventive measures regarding risk factor reduction. However, attention should be focused on the following key areas: identifying the initiation of disease, discovering a common link between known and emerging causal factors, exploring associations between suspected variables and the disease, and identifying prevention efforts.

Acknowledgments

The authors acknowledge the comments, suggestions, and insights provided by the scientific leadership of the Pulmonary Hypertension Association.

References

1. Rich S, Dantzker DR, Ayres SM, et al. Primary pulmonary hypertension: a national prospective study. Annals Intern Med 1987;107:216–23.
2. National Heart, Lung, and Blood Institute. Primary pulmonary hypertension. Bethesda, MD: US Department of Health and Human Services; November 1996. NIH publication no. 96-3291.
3. Gaine SP, Rubin LJ. Primary pulmonary hypertension. Lancet 1998;352:719–25.
4. Chatterjee K, De Marco T, Alpert JS. Pulmonary hypertension: hemodynamic diagnosis and management. Archives Intern Med 2002;162:1925–33.
5. D'Alonzo GE, Barst RJ, Ayres SM, et al. Survival in patients with primary pulmonary hypertension: results from a national prospective registry. Annals Intern Med 1991;115:343–9.
6. Kawut SM, Horn EM, Berekashvili KK, et al. New predictors of outcome in idiopathic pulmonary arterial hypertension. Am J Cardiol 2005;95:199–203.
7. Rubin LJ. ACCP consensus statement: primary pulmonary hypertension. Chest 1993;104:236–50.
8. McLaughlin VV, Rich S. Pulmonary hypertension—advances in medical and surgical interventions. J Heart Lung Transplant 1998;17:739–43.
9. Gaine SP, Rubin LJ. Medical and surgical treatment options for pulmonary hypertension. Am J Med Sci 1998;315:179–84.
10. Krowka MJ. Pulmonary hypertension: diagnostics and therapeutics. Mayo Clinic Proceedings 2000;75:625–30.
11. McLaughlin VV, Presberg KW, Doyle RL, et al. Prognosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. Chest 2004;126(Suppl 1):78S–92S.
12. Peacock AJ. Treatment of pulmonary hypertension: several options exist, but they are expensive and necessitate specialist care [Editorial]. Brit Med J 2003;326:835–6.
13. Fishman AP. Primary pulmonary arterial hypertension: a look back. J Am Coll Cardiol 2004;43:2S–4S.
14. Fishman AP. Clinical classification of pulmonary hypertension. Clinics in Chest Med 2001;22:385–91.
15. Lilienfeld DE, Rubin LJ. Mortality from primary pulmonary hypertension in the United States, 1979–1996. Chest 2000;117:796–800.
16. Rich S, Chomka E, Hasara L, et al. The prevalence of pulmonary hypertension in the United States: adult population estimates obtained from measurements of chest roentgenograms from the NHANES II survey. Chest 1989;96:236–41.
17. World Health Organization. Manual of the international statistical classification of diseases, injuries, and causes of death, 9th Revision, Vol. 1. Geneva, Switzerland: World Health Organization; 1977.
18. World Health Organization. International statistical classification of diseases and related health problems, 10th revision. Geneva, Switzerland: World Health Organization; 1992.
19. Anderson RN, Minino AM, Hoyert DL, Rosenberg HM. Comparability of cause of death between ICD-9 and ICD-10: preliminary estimates. Natl Vital Stat Rep 2001;49(2).
20. National Center for Health Statistics. National Vital Statistics System. Hyattsville, MD: US Department of Health and Human Services, CDC, National Center for Health Statistics; 2002. Available at http://www.cdc.gov/nchs/nvss.htm.
21. Anderson RN, Rosenberg HM. Age standardization of death rates: implementation of the year 2000 standard. Natl Vital Stat Rep 1998;47(3).
22. Klein RJ, Schoenborn CA. Age adjustment using the 2000 projected US population. Healthy People 2010 Statistics Notes, No. 20. Hyattsville, MD: US Department of Health and Human Services, CDC, National Center for Health Statistics; 2001.
23. Mannino DM, Homa DM, Akinbami LJ, Ford ES, Redd SC. Chronic obstructive pulmonary disease surveillance—United States, 1971–2000. In: Surveillance Summaries, August 2, 2002. MMWR 2002;51(No. SS-6):1–16.
24. Simmons WR, Schnack GA. Development of the design of the NCHS Hospital Discharge Survey. Vital Health Statistics, series 2, no. 39. Rockville, MD: Human Resources Administration; 1977. DHEW publication no. (HRA)77-1199. Available at http://www.cdc.gov/nchs/data/series/sr_02/sr02_039.pdf.
25. Dennison CF, Pokras R. Design and operation of the National Hospital Discharge Survey: 1988 redesign. Vital Health Stat 2000;1(39).
26. US Department of Health and Human Services. The International Classification of Diseases, Ninth Revision, Clinical Modification. 4th ed. Hyattsville, MD: US Department of Health and Human Services, CDC, National Center for Health Statistics, Health Care Financing Administration; 1991. DHHS publication no. (PHS) 91-1260.
27. Kozak LJ. Underreporting of race in the National Hospital Discharge Survey. Hyattsville, MD: US Department of Health and Human Services, CDC, National Center for Health Statistics; 1995. Advance data no. 265. Available at http://www.cdc.gov/nchs/data/ad/ad265.pdf.
28. Abenhaim L, Moride Y, Brenot F, et al. Appetite-suppressant drugs and the risk of primary pulmonary hypertension. N Engl J Med 1996;335:609–61.
29. Abenhaim L, Humbert M. Pulmonary hypertension related to drugs and toxins. Current Opinion in Cardiology 1999;14:437–41.
30. Rich S, Rubin L, Walker AM, Schneeweiss S, Abenhaim L. Anorexigens and pulmonary hypertension in the United States: results from the surveillance of North American pulmonary hypertension. Chest 2000;117:870–4.
31. Stafford RS, Radley DC. National trends in antiobesity medicare use. Arch Intern Med 2003;163:1046–50.

32. Khan LK, Serdula MK, Bowman BA, Williamson DF. Use of prescription weight loss pills among U.S. adults in 1996–1998. Ann Intern Med 2001;134:282–6.
33. Israel RA, Rosenberg HM, Curtin LR. Analytical potential for multiple cause-of-death data. Am J Epidemiol 1986;124:161–79.
34. Rich S. Clinical insights into the pathogenesis of primary pulmonary hypertension. Chest 1998;114:237S–241S.
35. Rubin LJ. Primary pulmonary hypertension. N Engl J Med 1997;336:111–7.
36. Archer S, Rich S. Primary pulmonary hypertension: a vascular biology and translational "work in progress." Circulation 2000;102:2781–91.
37. Hoepner MM, Galie N, Simonneau G, Rubin LJ. New treatments for pulmonary arterial hypertension. Am J Respir Crit Care Med 2002;165:1209–16. 2004;43(Suppl):5S–12S.
38. Simonneau G, Galie N, Rubin LJ, et al. Clinical classification of pulmonary hypertension. J Am Coll Cardiol 2004;43(Suppl):5S–12S.
39. Gaine S. Pulmonary hypertension. JAMA 2000;284:3160–8.
40. Barst RJ. Role of atrial septostomy in the treatment of pulmonary vascular disease. Thorax 2000;55:95–6.
41. Klepetko W, Mayer E, Sandoval J, et al. Interventional and surgical modalities of treatment of pulmonary arterial hypertension. J Am Coll Cardiol 2004;43(Suppl S):73S–80S.
42. Galie N, Seeger W, Naeije R, Simonneau G, Rubin LJ. Comparative analysis of clinical trials and evidence-based treatment algorithm in pulmonary arterial hypertension. J Am Coll Cardiol 2004;43 (Suppl S):81S–88S.
43. Barst RJ, McGoon M, Torbicki A, et al. Diagnosis and differential assessment of pulmonary arterial hypertension. J Am Coll Cardiol 2004;43(Suppl S):40S–47S.
44. Recusani R, DiMatteo A, Gambarin F, D'Armini A, Klersy C, Campana C. Clinical and therapeutical follow-up of HIV-associated pulmonary hypertension: prospective study of 10 patients. AIDS 2003;17 (Suppl 1):S88–S95.
45. Kiss H, Egarter C, Asseryanis E, Putz D, Kneussl M. Primary pulmonary hypertension in pregnancy: a case report. Am J Obstet Gynecol 1995;172:1052–4.
46. Easterling TR, Ralph DD, Schmucker BC. Pulmonary hypertension in pregnancy: treatment with pulmonary vasodilators. Obstet Gynecol 1999;93:494–8.
47. Monnery L, Nanson J, Charlton G. Primary pulmonary hypertension in pregnancy; a role for novel vasodilators. Br J Anaesth 2001;87:295–8.
48. Farber HW, Loscalzo J. Mechanisms of disease: pulmonary arterial hypertension. N Engl J Med 2004;351:1655–65.

TABLE 1. Number of decedents with pulmonary hypertension* as any contributing cause of death and age-standardized death rate,† by sex and year — United States, 1980–2002

Year	Men		Women		Total	
	No.	Rate	No.	Rate	No.	Rate
1980	6,999	8.2	3,923	3.3	**10,922**	**5.2**
1981	6,686	7.7	4,042	3.3	**10,728**	**5.0**
1982	6,306	7.2	3,910	3.2	**10,216**	**4.7**
1983	6,462	7.3	4,423	3.5	**10,885**	**5.0**
1984	6,655	7.4	4,673	3.7	**11,328**	**5.1**
1985	6,644	7.3	4,761	3.7	**11,405**	**5.1**
1986	6,350	6.9	4,946	3.8	**11,296**	**5.0**
1987	6,240	6.6	5,189	3.9	**11,429**	**5.0**
1988	6,302	6.6	5,386	4.0	**11,688**	**5.0**
1989	6,388	6.6	5,517	4.1	**11,905**	**5.0**
1990	6,391	6.5	5,689	4.1	**12,080**	**5.0**
1991	6,359	6.4	6,142	4.4	**12,501**	**5.1**
1992	6,317	6.2	6,417	4.5	**12,734**	**5.2**
1993	6,498	6.3	6,594	4.6	**13,092**	**5.2**
1994	6,206	5.9	6,766	4.6	**12,972**	**5.1**
1995	6,217	5.8	6,923	4.8	**13,140**	**5.1**
1996	6,317	5.9	7,186	4.8	**13,503**	**5.2**
1997	6,520	5.9	7,766	5.1	**14,286**	**5.4**
1998	6,431	5.8	8,042	5.2	**14,473**	**5.4**
1999	6,618	5.9	8,428	5.4	**15,046**	**5.5**
2000	6,273	5.5	8,629	5.4	**14,902**	**5.4**
2001	6,514	5.6	9,083	5.6	**15,597**	**5.5**
2002	6,484	5.4	9,184	5.5	**15,668**	**5.4**

* *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 during 1980–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 during 1999–2002.
† Per 100,000 population. Age-standardized to the 2000 U.S. standard population.

TABLE 2. Number of decedents with pulmonary hypertension* as any contributing cause of death and age-standardized death rate,† by race and year — United States, 1980–2002

Year	White		Black		Asian/ Pacific Islander		American Indian/ Alaska Native	
	No.	Rate	No.	Rate	No.	Rate	No.	Rate
1980	9,901	5.3	929	4.6	45	§	33	§
1981	9,699	5.1	944	4.6	49	§	24	§
1982	9,142	4.7	966	4.6	65	§	25	§
1983	9,725	5.0	1,052	5.0	59	§	34	§
1984	10,124	5.1	1,078	5.0	56	§	35	§
1985	10,112	5.1	1,151	5.4	64	§	40	§
1986	9,895	4.9	1,272	5.8	73	§	35	§
1987	10,054	4.9	1,223	5.5	93	§	35	§
1988	10,274	4.9	1,259	5.6	83	§	32	§
1989	10,395	4.9	1,353	5.9	114	§	43	§
1990	10,613	5.0	1,317	5.7	111	2.4	39	¶
1991	10,838	5.0	1,481	6.4	143	3.0	39	¶
1992	11,064	5.0	1,496	6.3	133	2.6	41	¶
1993	11,413	5.1	1,490	6.1	137	2.7	52	¶
1994	11,198	5.0	1,572	6.3	141	2.5	61	¶
1995	11,275	4.9	1,642	6.6	170	2.8	53	¶
1996	11,619	5.0	1,660	6.6	164	2.4	60	¶
1997	12,269	5.2	1,770	6.8	188	2.8	59	¶
1998	12,408	5.2	1,815	6.9	190	2.7	60	¶
1999	12,740	5.3	2,043	7.6	195	2.8	68	¶
2000	12,682	5.2	1,952	7.2	200	2.5	68	¶
2001	13,279	5.4	2,032	7.3	219	2.6	67	¶
2002	13,335	5.3	2,061	7.3	203	2.2	69	¶

* *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 during 1980–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 during 1999–2002.
† Per 100,000 population. Age-standardized to the 2000 U.S. standard population.
§ American Indian/Alaska Native and Asian/Pacific Islander categories did not exist in Census data before 1990.
¶ Numbers might be too small to calculate stable age-standardized death rates.

TABLE 3. Number of decedents with pulmonary hypertension* as any contributing cause of death and age-standardized death rate,† by Hispanic origin and year — United States, 1990–2002

	Hispanic		Non-Hispanic	
Year	No.	Rate	No.	Rate
1990	369	2.7	11,246	4.9
1991	419	3.3	11,806	5.1
1992	433	3.1	12,015	5.1
1993	401	2.9	12,486	5.2
1994	414	2.7	12,395	5.1
1995	471	3.1	12,490	5.1
1996	468	2.9	12,855	5.2
1997	538	3.2	13,697	5.5
1998	596	3.5	13,829	5.4
1999	552	3.0	14,447	5.6
2000	625	3.4	14,234	5.5
2001	645	3.2	14,916	5.6
2002	617	3.0	15,003	5.6

* *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 during 1990–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 during 1999–2002.
† Per 100,000 population. Age-standardized to the 2000 U.S. standard population.

TABLE 4. Number of decedents with pulmonary hypertension* as any contributing cause of death and age-specific death rate,† by age group and year — United States, 1980–2002

	0–44 yrs		45–54 yrs		55–64 yrs		65–74 yrs		75–84 yrs		≥85 yrs	
Year	No.	Rate	No.	Rate	No.	Rate	No.	Rate	No.	Rate	No.	Rate
1980	1,085	0.7	708	3.1	2,146	9.9	3,782	24.2	2,496	32.1	705	31.0
1981	1,065	0.7	675	3.0	2,112	9.6	3,820	24.0	2,407	30.2	649	27.6
1982	1,049	0.7	578	2.6	1,951	8.9	3,546	22.0	2,394	29.2	695	28.5
1983	1,189	0.7	598	2.7	2,068	9.4	3,670	22.4	2,650	31.4	709	28.1
1984	1,284	0.8	596	2.7	2,018	9.1	3,813	22.9	2,841	32.8	776	29.9
1985	1,294	0.8	629	2.8	2,020	9.1	3,693	21.9	2,908	32.7	860	32.2
1986	1,321	0.8	592	2.6	1,907	8.7	3,702	21.6	2,953	32.3	818	29.8
1987	1,325	0.8	600	2.6	1,893	8.7	3,648	20.9	3,101	33.1	860	30.5
1988	1,330	0.8	640	2.7	1,915	8.9	3,810	21.6	3,103	32.3	890	30.8
1989	1,471	0.9	626	2.5	1,884	8.9	3,731	20.9	3,271	33.2	921	31.0
1990	1,461	0.8	592	2.3	1,807	8.6	3,812	21.1	3,393	33.7	1,015	33.2
1991	1,465	0.8	653	2.5	1,724	8.2	3,849	21.1	3,662	35.5	1,147	36.0
1992	1,483	0.9	702	2.6	1,766	8.4	3,854	20.9	3,719	35.3	1,210	36.5
1993	1,460	0.8	666	2.3	1,784	8.5	3,897	20.9	3,965	36.9	1,320	38.3
1994	1,469	0.8	708	2.4	1,685	8.0	4,006	21.4	3,766	34.4	1,338	37.6
1995	1,584	0.9	773	2.5	1,710	8.1	3,662	19.5	3,894	34.8	1,517	41.2
1996	1,425	0.8	868	2.7	1,632	7.6	3,725	19.9	4,131	36.0	1,720	45.3
1997	1,611	0.9	919	2.7	1,724	7.9	3,744	20.2	4,348	37.0	1,939	49.5
1998	1,611	0.9	974	2.8	1,699	7.5	3,726	20.3	4,346	36.4	2,117	52.2
1999	1,630	0.9	1,014	2.8	1,779	7.6	3,677	20.2	4,519	37.2	2,427	58.1
2000	1,535	0.8	1,047	2.8	1,726	7.1	3,511	19.1	4,522	36.6	2,561	60.4
2001	1,635	0.9	1,081	2.8	1,793	7.1	3,503	19.1	4,755	37.8	2,829	64.2
2002	1,555	0.8	1,122	2.8	1,848	6.9	3,343	18.3	4,830	37.9	2,969	64.6

* *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 during 1980–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 during 1999–2002.
† Per 100,000 population.

TABLE 5. Age-standardized and age-specific death rates* for pulmonary hypertension† as any contributing cause of death for groups defined by selected characteristics, by period — United States, 1980–2002

Characteristic	1980–1984	1985–1989	1990–1994	1995–1999	2000–2002
Age-standardized§ death rate					
Total	5.0	5.0	5.1	5.3	5.4
Men	7.5	6.8	6.3	5.9	5.5
Women	3.4	3.9	4.5	5.1	5.5
Race					
White	5.0	4.9	5.0	5.1	5.3
Black	4.8	5.6	6.2	6.9	7.3
Asian/Pacific Islander	¶	¶	2.6	2.7	2.4
American Indian/Alaska Native	¶	¶	**	**	**
Hispanic origin					
Hispanic	¶	¶	2.9	3.1	3.2
Non-Hispanic	¶	¶	5.1	5.4	5.6
Age-specific death rate					
Total					
0–44 yrs	0.7	0.8	0.8	0.9	0.9
45–54 yrs	2.8	2.6	2.4	2.7	2.8
55–64 yrs	9.4	8.9	8.4	7.7	7.0
65–74 yrs	23.1	21.4	21.1	20.0	18.8
75–84 yrs	31.1	32.7	35.2	36.3	37.4
≥85 yrs	29.0	30.9	36.4	49.5	63.2
Men					
0–44 yrs	0.7	0.8	0.8	0.8	0.8
45–54 yrs	2.9	2.7	2.2	2.4	2.4
55–64 yrs	12.0	10.3	8.9	7.6	6.4
65–74 yrs	34.7	28.3	25.3	21.6	19.0
75–84 yrs	56.1	51.3	48.3	43.1	39.7
≥85 yrs	55.7	53.6	55.0	62.7	68.9
Women					
0–44 yrs	0.7	0.8	0.9	1.0	0.9
45–54 yrs	2.7	2.6	2.6	3.0	3.1
55–64 yrs	7.1	7.6	7.9	7.8	7.6
65–74 yrs	14.2	16.0	17.8	18.7	18.7
75–84 yrs	16.5	21.7	27.1	31.9	36.0
≥85 yrs	17.9	21.9	29.2	44.1	60.7
White					
0–44 yrs	0.6	0.7	0.7	0.7	0.7
45–54 yrs	2.6	2.4	2.1	2.3	2.4
55–64 yrs	9.3	8.7	8.1	7.2	6.6
65–74 yrs	23.8	21.7	21.1	19.8	18.6
75–84 yrs	32.2	33.5	35.8	36.6	37.9
≥85 yrs	30.0	31.3	37.0	50.5	64.1
Black					
0–44 yrs	1.3	1.6	1.6	1.7	1.7
45–54 yrs	4.8	5.3	5.4	6.2	6.2
55–64 yrs	11.2	11.8	11.4	12.8	12.5
65–74 yrs	17.4	20.5	23.5	23.8	25.2
75–84 yrs	20.1	25.4	30.6	35.6	38.7
≥85 yrs	17.5	25.9	29.9	43.6	56.1

* Per 100,000 population.

† *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 for deaths during 1980–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 for deaths during 1999–2002.

§ To the 2000 U.S. standard population.

¶ Hispanic origin and American Indian/Alaska Native and Asian/Pacific Islander race categories did not exist in Census data before 1990.

** Numbers might be too small to calculate stable age-standardized death rates.

TABLE 6. Annual number of decedents with pulmonary hypertension* as any contributing cause of death, aggregated number, and age-standardized death rate,† by state/area — United States, 2000–2002

	2000	2001	2002	2000–2002	
State/Area	No.	No.	No.	No.	Rate
Alabama	224	233	229	686	5.0
Alaska	26	22	30	78	7.7§
Arizona	192	229	216	637	4.0
Arkansas	103	99	105	307	3.6
California	1,432	1,488	1,423	4,343	4.8
Colorado	284	323	322	929	8.6
Connecticut	184	175	155	514	4.5
Delaware	48	65	53	166	6.8
District of Columbia	53	53	44	150	8.8
Florida	888	898	958	2,744	4.4
Georgia	335	375	312	1,022	4.9
Hawaii	52	51	60	163	4.3
Idaho	100	115	109	324	8.9
Illinois	646	666	688	2,000	5.5
Indiana	372	371	356	1,099	6.0
Iowa	196	225	216	637	6.1
Kansas	147	178	189	514	6.0
Kentucky	259	226	235	720	5.9
Louisiana	163	180	186	529	4.2
Maine	100	95	85	280	6.3
Maryland	312	298	293	903	6.0
Massachusetts	274	254	290	818	4.0
Michigan	593	565	609	1,767	6.0
Minnesota	302	303	385	990	6.6
Mississippi	123	153	141	417	5.0
Missouri	380	387	370	1137	6.3
Montana	59	87	72	218	7.4
Nebraska	99	104	102	305	5.4
Nevada	75	82	88	245	4.5
New Hampshire	87	97	80	264	7.2
New Jersey	392	385	391	1,168	4.4
New Mexico	104	114	135	353	6.8
New York	795	830	855	2,480	4.2
North Carolina	555	617	579	1,751	7.4
North Dakota	37	38	40	115	5.2
Ohio	853	908	896	2,657	7.4
Oklahoma	186	170	189	545	5.0
Oregon	219	230	243	692	6.4
Pennsylvania	844	876	854	2,574	5.8
Rhode Island	54	66	63	183	5.0
South Carolina	194	232	234	660	5.6
South Dakota	39	45	44	128	4.9
Tennessee	303	307	356	966	5.6
Texas	863	924	936	2,723	5.1
Utah	87	121	121	329	6.6
Vermont	46	53	62	161	8.4
Virginia	416	361	410	1,187	6.0
Washington	325	369	359	1,053	6.3
West Virginia	152	155	151	458	7.3
Wisconsin	288	345	304	937	5.5
Wyoming	42	54	45	141	9.9
Total	**14,902**	**15,597**	**15,668**	**46,167**	**5.4**

* *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 reported as any contributing cause of death on the death certificate
† Per 100,000 population. Age-standardized to the 2000 U.S. standard population.
§ Number of deaths might be too small to calculate a stable age-standardized death rate.

TABLE 7. Percentage of selected characteristics among decedents with pulmonary hypertension* reported as any contributing cause of death, by period — United States, 1980–2002

Characteristic	1980–1984 (N = 54,079)	1985–1989 (N = 57,723)	1990–1994 (N = 63,379)	1995–1999 (N = 70,448)	2000–2002 (N = 46,167)
Women	38.8%	44.7%	49.9%	54.4%	58.3%
Race					
White	89.9%	87.9%	87.0%	85.6%	85.1%
Black	9.2%	10.8%	11.6%	12.7%	13.1%
Other race/missing race	0.9%	1.3%	1.4%	1.7%	1.8%
Hispanic origin	†	†	3.2%	3.7%	4.1%
Age group (yrs)					
0–44	10.5%	11.7%	11.6%	11.2%	10.2%
45–54	5.8%	5.3%	5.2%	6.5%	7.0%
55–64	19.0%	16.7%	13.8%	12.1%	11.6%
65–74	34.5%	32.2%	30.6%	26.3%	22.4%
75–84	23.7%	26.6%	29.2%	30.1%	30.6%
≥85	6.5%	7.5%	9.5%	13.8%	18.1%

* *International Classification of Diseases, Ninth Revision* codes 416.0, 416.8, or 416.9 for deaths during 1980–1998 and *International Classification of Diseases, Tenth Revision* codes I27.0, I27.8, or I27.9 for deaths during 1999–2002.
† Information on Hispanic origin not available.

TABLE 8. Percentage of selected causes of death reported as the underlying cause of death among all decedents with pulmonary hypertension reported as any contributing cause of death, by period — United States, 1980–2002

Underlying cause of death reported on death certificate (ICD-9/ICD-10 codes*)	1980–1984 (N = 54,079)	1985–1989 (N = 57,723)	1990–1994 (N = 63,379)	1995–1999 (N = 70,448)	2000–2002 (N = 46,167)
Pulmonary hypertension (416.0, 416.8, 416.9/I27.0, I27.8, I27.9)	14.3%	17.6%	19.3%	23.5%	30.3%
Chronic lower respiratory diseases (490–494, 496/J40–J47)	49.2%	44.5%	40.8%	33.8%	25.9%
Coronary heart disease (410–414, 429.2/I20–I25)	8.4%	7.9%	7.6%	8.2%	8.6%
All other cardiovascular diseases (390–405, 415, 416.1, 417, 420–423, 425–427, 429.0, 429.1, 429.3–429.9, 430–434, 436–448/I00–I19, I26, I27.1, I28–I34, I40–I49, I51–I78)	5.2%	5.9%	6.6%	7.3%	7.2%
All other respiratory diseases (460–478, 495, 500–519/J00–J06, J20–J22, J60–J99)	6.6%	6.0%	6.0%	5.8%	5.9%
Congenital malformations, deformations, and chromosomal abnormalities (740–759/Q00–Q99)	2.9%	3.2%	3.4%	3.3%	2.9%
Chronic valvular heart disease (424/I35–I39)	0.5%	0.7%	1.3%	2.1%	2.8%
Malignant neoplasms of trachea, bronchus, and lung (162/C33–C44)	2.0%	2.0%	2.2%	2.0%	1.7%
Systemic lupus erythematosus (710.0/M32), systemic sclerosis (710.1/M34), dermatomyositis (710.3–710.4/M33), sicca syndrome (710.2/M35), rheumatoid arthritis (714.0–714.2/M05–M06), and juvenile arthritis (714.3/M08–M09)	0.8%	1.2%	1.7%	2.2%	1.2%
Complications of pregnancy, childbirth, and the puerperium (640–676/O10–O99), or conditions originating in the perinatal period (760–771.2, 771.4–779/P00–P96)	1.8%	2.3%	1.5%	1.2%	1.1%
Influenza and pneumonia (480–487/J10–J18)	1.4%	1.5%	1.4%	1.3%	1.0%
Heart failure (428/I50)	0.7%	0.8%	0.8%	0.9%	0.7%
Chronic liver disease and cirrhosis (571/K70, K73–K74)	0.2%	0.3%	0.3%	0.3%	0.3%
Human immunodeficiency virus infection (042–044/B20–B24)	0	0	0.1%	0.1%	0.1%
All other causes	6.0%	6.1%	7.0%	8.1%	10.2%

* *International Classification of Diseases, Ninth Revision* (ICD-9) codes 416.0, 416.8, or 416.9 for deaths during 1980–1998 and *International Classification of Diseases, Tenth Revision* (ICD-10) codes I27.0, I27.8, or I27.9 for deaths during 1999–2002.

TABLE 9. Percentage of selected causes of death reported as the underlying cause of death among decedents aged <45 years with pulmonary hypertension reported as any contributing cause of death, by period — United States, 1980–2002

Underlying cause of death reported on death certificate (ICD-9/ICD-10 codes*)	1980–1984 (N = 5,672)	1985–1989 (N = 6,741)	1990–1994 (N = 7,338)	1995–1999 (N = 7,861)	2000–2002 (N = 4,725)
Pulmonary hypertension (416.0, 416.8, 416.9/I27.0, I27.8, I27.9)	29.8%	32.7%	36.4%	39.1%	39.8%
Congenital malformations, deformations, and chromosomal abnormalities (740–759/Q00–Q99)	20.9%	21.4%	23.0%	22.3%	21.7%
Complications of pregnancy, childbirth, and the puerperium (640–676/O10–O99), or conditions originating in the perinatal period (760–771.2, 771.4–779/P00–P96)	17.3%	19.6%	12.8%	10.3%	10.5%
All cardiovascular diseases, excluding pulmonary hypertension (390–415, 416.1, 417–448/I00–I26, I27.1, I28–I78)	5.4%	5.6%	6.3%	6.8%	6.2%
All other respiratory diseases (460–478, 495, 500–519/J00–J06, J20–J22, J60–J99)	4.1%	3.5%	3.4%	3.4%	4.2%
Chronic lower respiratory diseases (490–494, 496/J40–J47)	5.2%	3.2%	2.6%	2.5%	2.7%
Systemic lupus erythematosus (710.0/M32), systemic sclerosis (710.1/M34), dermatomyositis (710.3–710.4/M33), sicca syndrome (710.2/M35), rheumatoid arthritis (714.0–714.2/M05–M06), and juvenile arthritis (714.3/M08–M09)	2.1%	2.7%	3.6%	3.7%	2.4%
Influenza and pneumonia (480–487/J10–J18)	1.1%	1.0%	0.9%	1.0%	0.6%
All other causes	14.1%	10.3%	11.0%	10.9%	11.9%

* *International Classification of Diseases, Ninth Revision* (ICD-9) codes 416.0, 416.8, or 416.9 for deaths during 1980–1998 and *International Classification of Diseases, Tenth Revision* (ICD–10) codes I27.0, I27.8, or I27.9 for deaths during 1999–2002.

TABLE 10. Estimated annual number of hospitalizations and age-standardized hospital rate* for persons with pulmonary hypertension† as any-listed diagnosis during hospital stay, by year and sex — National Hospital Discharge Survey, United States, 1980–2002

	Men		Women		Total	
Year	No.	Rate	No.	Rate	No.	Rate
1980	47,000	52.0	40,000	33.5	**87,000**	**40.8**
1981	51,000	55.5	34,000	28.0	**85,000**	**39.1**
1982	62,000	67.2	39,000	31.9	**101,000**	**46.1**
1983	58,000	61.8	45,000	35.8	**103,000**	**46.1**
1984	67,000	68.1	56,000	45.0	**123,000**	**54.5**
1985	57,000	57.8	54,000	41.6	**111,000**	**48.1**
1986	60,000	61.7	60,000	46.9	**120,000**	**52.1**
1987	59,000	58.4	49,000	37.5	**108,000**	**45.9**
1988	56,000	54.4	58,000	43.8	**114,000**	**47.8**
1989	56,000	52.4	53,000	40.7	**109,000**	**45.5**
1990	55,000	53.0	61,000	45.1	**116,000**	**48.4**
1991	69,000	64.1	71,000	53.9	**140,000**	**58.5**
1992	68,000	62.6	83,000	60.6	**151,000**	**61.6**
1993	67,000	62.6	87,000	61.8	**154,000**	**61.9**
1994	79,000	75.8	84,000	58.1	**163,000**	**64.9**
1995	83,000	77.4	101,000	69.8	**184,000**	**71.9**
1996	76,000	68.9	111,000	76.2	**187,000**	**72.7**
1997	86,000	76.6	120,000	80.9	**206,000**	**78.6**
1998	105,000	92.3	146,000	97.5	**251,000**	**94.4**
1999	110,000	95.5	146,000	94.8	**256,000**	**94.6**
2000	113,000	97.6	172,000	110.1	**285,000**	**104.1**
2001	102,000	84.3	160,000	99.2	**262,000**	**92.7**
2002	103,000	82.3	157,000	95.3	**260,000**	**90.1**

* Per 100,000 population. Age-standardized to the 2000 U.S. standard population.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for hospital diagnoses during 1980–2002.

TABLE 11. Estimated annual number* of hospitalizations and age-specific hospital rate† for persons with pulmonary hypertension§ as any-listed diagnosis during hospital stay, by year and age group — National Hospital Discharge Survey, United States, 1980–2002

	0–44 yrs		45–54 yrs		55–64 yrs		65–74 yrs		75–84 yrs		≥85 yrs	
Year	No.	Rate	No.	Rate	No.	Rate	No.	Rate	No.	Rate	No.	Rate
1980	10,000	6.4	8,000	33.1	20,000	94.2	31,000	198.4	15,000	191.9	3,000	146.9
1981	11,000	6.9	5,000	25.1	21,000	95.8	30,000	189.3	15,000	188.8	3,000	122.6
1982	13,000	8.2	10,000	43.0	24,000	109.9	33,000	205.9	18,000	226.1	3,000	106.4
1983	10,000	6.3	7,000	29.2	28,000	127.2	34,000	209.5	21,000	251.4	3,000	117.5
1984	14,000	8.8	11,000	49.4	31,000	138.2	41,000	245.4	22,000	253.0	4,000	159.5
1985	12,000	7.4	7,000	32.8	27,000	121.6	42,000	247.1	18,000	201.5	5,000	184.6
1986	17,000	10.0	10,000	42.8	22,000	100.7	41,000	241.2	26,000	281.9	4,000	156.0
1987	12,000	7.2	5,000	23.0	24,000	111.1	38,000	218.5	24,000	256.4	4,000	156.9
1988	17,000	10.5	7,000	29.5	28,000	128.4	37,000	211.4	19,000	196.4	5,000	193.5
1989	21,000	12.4	10,000	39.6	21,000	98.0	33,000	183.8	20,000	208.3	4,000	131.8
1990	14,000	8.1	12,000	47.2	20,000	97.3	42,000	235.1	23,000	234.0	4,000	146.4
1991	17,000	9.4	19,000	72.5	26,000	124.9	43,000	240.6	28,000	274.0	7,000	227.3
1992	18,000	10.2	14,000	50.6	26,000	125.4	47,000	259.6	37,000	357.3	8,000	261.6
1993	17,000	9.3	13,000	46.0	25,000	119.9	49,000	263.8	40,000	377.9	10,000	312.6
1994	14,000	7.5	13,000	43.7	22,000	107.2	52,000	283.1	46,000	424.9	16,000	450.1
1995	21,000	11.8	17,000	54.7	24,000	112.7	56,000	302.0	47,000	421.9	19,000	527.1
1996	20,000	10.8	20,000	62.2	28,000	133.5	51,000	276.5	49,000	436.8	19,000	503.3
1997	22,000	11.9	21,000	62.9	28,000	129.0	53,000	288.9	63,000	543.7	19,000	491.8
1998	21,000	11.4	24,000	69.1	43,000	192.0	67,000	369.3	69,000	580.8	27,000	676.2
1999	22,000	12.2	21,000	58.1	37,000	161.2	71,000	394.6	73,000	608.0	31,000	740.6
2000	22,000	12.3	23,000	60.6	38,000	160.1	72,000	400.9	87,000	713.7	43,000	997.6
2001	25,000	13.7	23,000	57.7	47,000	187.5	57,000	312.9	75,000	595.8	35,000	793.9
2002	28,000	15.2	24,000	58.7	45,000	169.4	58,000	315.8	65,000	512.1	40,000	880.3

* Estimated annual numbers for each age group are rounded to the nearest thousand; the sum of these numbers might not equal the total estimated annual number of hospitalizations in Table 10.
† Per 100,000 population.
§ *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for hospital diagnoses during 1980–2002.

TABLE 12. Estimated age-standardized and age-specific hospital rates* for groups with pulmonary hypertension† as any-listed diagnosis, by selected characteristics and period — National Hospital Discharge Survey, United States, 1980–2002

Characteristic	1980–1984	1985–1989	1990–1994	1995–1999	2000–2002
Age-standardized§ hospital rate					
Total	45.4	47.8	59.2	82.7	95.5
Men	61.0	56.9	63.9	82.4	87.9
Women	34.9	42.1	56.0	84.0	101.5
Age-specific hospital rate					
Total					
0–44 yrs	7.3	9.5	8.9	11.6	13.7
45–54 yrs	35.9	33.6	51.6	61.5	59.0
55–64 yrs	113.2	112.0	114.9	146.5	172.5
65–74 yrs	210.1	220.0	256.7	325.9	342.9
75–84 yrs	223.2	228.5	335.6	520.5	605.8
≥85 yrs	130.7	164.2	284.6	592.3	889.3
Men					
0–44 yrs	5.9	8.6	9.2	9.0	12.5
45–54 yrs	34.7	29.9	46.3	58.2	52.9
55–64 yrs	139.3	127.8	118.8	149.1	154.5
65–74 yrs	311.1	277.5	291.5	328.7	308.4
75–84 yrs	378.4	308.4	389.0	507.4	600.8
≥85 yrs	182.5	257.8	294.8	723.1	766.2
Women					
0–44 yrs	8.8	10.5	8.6	14.3	15.0
45–54 yrs	37.1	37.0	56.7	64.6	64.8
55–64 yrs	90.1	98.0	111.4	144.2	189.2
65–74 yrs	132.6	175.3	229.1	323.5	371.6
75–84 yrs	131.9	181.3	302.7	529.2	609.0
≥85 yrs	109.1	127.3	280.7	538.8	942.1

* Per 100,000 population.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for hospital diagnoses during 1980–2002.
§ To the 2000 U.S. standard population.

TABLE 13. Percentage of persons with selected characteristics hospitalized with pulmonary hypertension* as any-listed diagnosis, by period — United States, 1980–2002

Characteristic	1980–1984 (N = 500,000)	1985–1989 (N = 561,000)	1990–1994 (N = 725,000)	1995–1999 (N = 1,083,000)	2000–2002 (N = 807,000)
Women	42.6%	48.9%	53.2%	57.6%	60.6%
Age group (yrs)					
0–44	11.6%	14.1%	10.9%	9.7%	9.4%
45–54	8.1%	7.0%	9.8%	9.5%	8.5%
55–64	24.9%	21.7%	16.5%	14.8%	16.2%
65–74	33.9%	34.1%	32.3%	27.6%	23.2%
75–84	18.3%	19.1%	24.2%	27.8%	28.2%
≥85	3.2%	4.1%	6.4%	10.6%	14.6%

* *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for hospital diagnoses during 1980–2002.

TABLE 14. Percentage of selected diseases reported as the principal (i.e., first-listed) diagnosis on hospital records with pulmonary hypertension reported as any-listed diagnosis, by period — United States, 1980–2002

Principal diagnosis reported on hospital record (ICD-9-CM codes*)	1980–1984 (N = 500,000)	1985–1989 (N = 561,000)	1990–1994 (N = 725,000)	1995–1999 (N = 1,083,000)	2000–2002 (N = 807,000)
Heart failure (428)	5.9%	11.4%	14.6%	17.7%	18.7%
All other cardiovascular diseases (390–405, 415, 416.1,420–423,425–427,429.0, 429.1,429.3–429.9,430–434, 436–448)	6.8%	9.6%	11.8%	14.9%	16.3%
Chronic lower respiratory diseases (490–494, 496)	42.0%	23.1%	16.4%	13.4%	12.9%
Coronary heart disease (410–414, 429.2)	3.2%	5.0%	7.9%	9.2%	8.6%
Influenza and pneumonia (480–487)	4.5%	8.0%	7.7%	6.5%	8.4%
All other respiratory diseases (460–478, 495, 500–519)	6.7%	13.3%	11.2%	9.2%	6.5%
Pulmonary hypertension (416.0, 416.8, 416.9)	12.8%	10.7%	6.9%	5.3%	4.2%
Chronic valvular heart disease (424)	1.0%	1.3%	1.9%	2.3%	1.9%
Congenital malformations, deformations, and chromosomal abnormalities (740–759)	3.0%	2.6%	2.9%	1.4%	0.7%
Chronic liver disease and cirrhosis (571)	0.2%	0.1%	0.2%	0.5%	0.5%
Complications of pregnancy, childbirth, and the puerperium (640–676), or conditions originating in the perinatal period (760–771.2, 771.4–779)	0.7%	1.8%	1.3%	0.2%	0.3%
Malignant neoplasms of trachea, bronchus, and lung (162)	0.7%	0.4%	0.4%	0.6%	0.2%
Systemic lupus erythematosus (710.0), systemic sclerosis (710.1), dermatomyositis (710.3–710.4), sicca syndrome (710.2), rheumatoid arthritis (714.0–714.2), and juvenile arthritis (714.3)	0.7%	0.2%	0.7%	0.3%	0.1%
Human immunodeficiency virus infection (042–044)	0	0	0	0.1%	0
All other causes	11.8%	12.6%	16.2%	18.7%	20.7%

**International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for hospital diagnoses during 1980–2002.

TABLE 15. Percentage of selected diseases reported as the principal (i.e., first-listed) diagnosis on hospital records of persons aged <45 years with pulmonary hypertension as any-listed diagnosis, by period — United States, 1980–2002

Principal diagnosis reported on the hospital record (ICD-9-CM codes*)	1980–1984 (N = 58,000)	1985–1989 (N = 79,000)	1990–1994 (N = 79,000)	1995–1999 (N = 105,000)	2000–2002 (N = 76,000)
All other cardiovascular diseases, excluding pulmonary hypertension (390–415, 416.1, 419–448)	13.9%	16.6%	19.9%	28.3%	29.8%
All other respiratory diseases (460–478, 495, 500–519)	3.5%	11.2%	10.0%	9.0%	9.8%
Pulmonary hypertension (416.0, 416.8, 416.9)	26.0%	15.9%	9.7%	9.5%	9.5%
Chronic lower respiratory diseases (490–494, 496)	5.2%	4.6%	3.7%	4.3%	7.1%
Influenza and pneumonia (480–487)	3.5%	6.4%	7.4%	7.2%	4.6%
Congenital malformations, deformations, and chromosomal abnormalities (740–759)	22.9%	16.1%	23.6%	12.7%	4.3%
Complications peculiar to certain specific procedures (996)	0.3%	2.2%	0.9%	2.2%	3.9%
Cellulitis and abscess (681–682)	0	0	0.1%	1.0%	3.6%
Complications of pregnancy, childbirth, and the puerperium (640–676), or conditions originating in the perinatal period (760–771.2, 771.4–779)	5.9%	12.2%	11.8%	2.4%	3.2%
Systemic lupus erythematosus (710.0), systemic sclerosis (710.1), dermatomyositis (710.3–710.4), sicca syndrome (710.2), rheumatoid arthritis (714.0–714.2), and juvenile arthritis (714.3)	3.1%	0.7%	0.3%	1.1%	0.7%
All other causes	15.7%	14.1%	12.6%	22.3%	23.5%

* *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed hospital diagnoses during 1980–2002.

TABLE 16. Annual number of Medicare hospitalizations and age-standardized hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension† as any-listed diagnosis during hospital stay, by sex and year — United States, 1990–2002

	Men		Women		Total	
Year	No.	Rate	No.	Rate	No.	Rate
1990	26,875	244.4	28,641	169.7	**55,516**	**197.8**
1991	29,229	262.5	32,959	192.5	**62,188**	**218.8**
1992	38,030	338.7	44,330	256.2	**82,360**	**287.1**
1993	43,142	381.4	51,977	298.0	**95,119**	**329.3**
1994	50,600	446.8	62,703	359.5	**113,303**	**392.4**
1995	55,575	493.9	72,318	417.3	**127,893**	**446.1**
1996	60,946	549.7	83,055	486.5	**144,001**	**510.3**
1997	67,405	625.0	95,125	569.1	**162,530**	**589.8**
1998	69,454	658.9	102,081	624.9	**171,535**	**636.8**
1999	72,028	686.9	108,667	671.0	**180,695**	**675.8**
2000	73,292	684.1	115,824	706.8	**189,116**	**696.3**
2001	69,300	622.3	112,074	666.8	**181,374**	**648.1**
2002	70,123	607.4	117,082	679.4	**187,205**	**649.7**

* Per 100,000 Medicare enrollees. Age-standardized to 2000 U.S. standard population aged ≥65 years.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed diagnosis on Medicare hospital claims during 1990–2002.

TABLE 17. Annual number of Medicare hospitalizations and age-standardized hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension† as any-listed diagnosis during hospital stay, by race and year — United States, 1990–2002

	White		Black		Asian		Hispanic		American Indian	
Year	No.	Rate	No.	Rate	No.	Rate	No.	Rate	No.	Rate
1990	48,610	198.1	4,381	210.2	§	§	§	§	§	§
1991	54,278	219.5	4,951	233.0	§	§	§	§	§	§
1992	71,498	287.5	6,625	309.1	§	§	§	§	§	§
1993	82,316	329.7	7,840	363.3	§	§	§	§	§	§
1994	99,765	389.0	10,112	445.7	119	—¶	346	—¶	36	—¶
1995	112,856	445.1	11,688	523.2	276	—¶	732	—¶	96	—¶
1996	126,480	507.4	13,413	613.6	336	—¶	906	—¶	121	—¶
1997	142,504	587.4	15,235	717.3	536	—¶	1,540	329.9	160	—¶
1998	149,908	633.1	16,459	796.8	729	—¶	1,992	439.8	198	—¶
1999	157,691	671.8	17,138	838.6	809	—¶	2,197	495.3	215	—¶
2000	164,692	687.8	18,293	868.6	765	—¶	2,294	471.6	219	—¶
2001	157,857	641.0	17,703	808.9	909	—¶	2,077	403.8	250	—¶
2002	162,179	641.2	18,572	823.0	982	—¶	2,352	437.2	503	—¶

* Per 100,000 Medicare enrollees. Age-standardized to 2000 U.S. standard population aged ≥65 years.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed diagnosis on Medicare hospital claims during 1990–2002.
§ Information was not available for specific race or ethnic groups other than white or black.
¶ Number of hospitalizations might be too small to calculate stable age-standardized hospital rate.

TABLE 18. Annual number of Medicare hospitalizations and age-specific hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension† as any-listed diagnosis during hospital stay, by year and age group — United States, 1990–2002

	65–74 yrs		75–84 yrs		≥85 yrs	
Year	No.	Rate	No.	Rate	No.	Rate
1990	30,296	187.8	20,635	228.0	4,585	151.4
1991	32,713	201.0	23,622	255.2	5,853	187.8
1992	41,642	254.1	32,040	341.4	8,678	270.3
1993	46,294	282.1	37,973	399.1	10,852	328.6
1994	53,706	329.3	45,475	475.7	14,122	420.8
1995	58,161	364.0	51,959	540.6	17,773	522.4
1996	62,295	403.7	59,666	620.9	22,040	644.0
1997	64,683	438.7	68,855	722.2	28,992	846.6
1998	64,944	460.6	74,160	788.0	32,431	943.7
1999	65,374	476.1	78,312	830.6	37,009	1,067.9
2000	65,862	474.8	82,355	858.1	40,899	1,157.4
2001	62,955	440.1	78,294	791.1	40,125	1,106.8
2002	63,557	431.9	80,763	791.1	42,885	1,154.2

* Per 100,000 Medicare enrollees.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed diagnosis on Medicare hospital claims during 1990–2002.

TABLE 19. Age-standardized and age-specific Medicare hospital rates* for pulmonary hypertension† as any-listed diagnosis for groups defined by selected characteristics, by period — United States, 1990–2002

Characteristic	1990–1994	1995–1999	2000–2002
Age-standardized§ hospital rate			
Total	286.2	570.3	664.2
Men	336.2	602.2	637.0
Women	256.1	551.7	684.1
Race			
White	286.1	567.3	656.3
Black	314.8	694.4	832.9
Asian	¶	314.8	281.1
Hispanic	¶	407.3	437.2
American Indian	¶	592.4	612.2
Age-specific hospital rate			
Total			
65–74 yrs	251.1	426.4	448.5
75–84 yrs	341.6	699.6	812.7
≥85 yrs	275.3	806.0	1,139.4
Men			
65–74 yrs	274.6	433.4	419.7
75–84 yrs	415.2	751.9	791.5
≥85 yrs	356.6	923.8	1,190.3
Women			
65–74 yrs	232.3	420.6	473.1
75–84 yrs	297.1	666.7	826.6
≥85 yrs	245.0	761.7	1,119.2
White			
65–74 yrs	250.6	418.8	430.4
75–84 yrs	341.9	700.0	811.0
≥85 yrs	275.3	814.1	1,157.1
Black			
65–74 yrs	287.2	594.4	665.1
75–84 yrs	360.7	796.8	976.2
≥85 yrs	303.0	821.8	1,117.4

* Per 100,000 Medicare enrollees.
† *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed diagnosis on Medicare hospital claims during 1990–2002.
§ To 2000 U.S. standard population aged ≥65 years.
¶ Information was not available for specific race or ethnic populations other than white or black.

TABLE 20. Annual number of Medicare hospitalizations with pulmonary hypertension* as any-listed diagnosis, aggregated number, and age-standardized hospital rate,† by state/area — United States, 2000–2002

	2000	2001	2002	2000–2002	
State/Area	No.	No.	No.	No.	Rate
Alabama	3,859	3,452	3,582	10,893	720.9
Alaska	209	191	243	643	659.9
Arizona	1,974	2,066	2,368	6,408	554.1
Arkansas	1,373	1,425	1,507	4,305	417.0
California	9,086	8,839	9,465	27,390	474.9
Colorado	2,703	2,610	2,679	7,992	1,022.8
Connecticut	1,856	1,898	2,067	5,821	484.4
Delaware	686	812	708	2,206	781.7
District of Columbia	508	447	407	1,362	782.0
Florida	14,688	13,378	14,185	42,251	732.1
Georgia	4,631	4,414	4,590	13,635	648.7
Hawaii	202	207	206	615	218.9
Idaho	1,121	1,036	974	3,131	796.3
Illinois	9,902	9,960	10,123	29,985	773.9
Indiana	5,610	5,040	5,203	15,853	748.5
Iowa	2,982	2,790	2,823	8,595	686.8
Kansas	1,929	1,791	2,052	5,772	594.3
Kentucky	3,334	3,173	3,119	9,626	703.1
Louisiana	2,726	2,484	2,691	7,901	638.4
Maine	1,172	1,043	1,145	3,360	626.0
Maryland	4,237	4,649	4,578	13,464	841.1
Massachusetts	3,694	3,653	3,863	11,210	591.8
Michigan	7,606	7,390	8,030	23,026	671.0
Minnesota	3,634	3,378	3,304	10,316	690.9
Mississippi	2,024	1,793	1,980	5,797	599.9
Missouri	4,312	4,384	4,470	13,166	713.5
Montana	998	998	975	2,971	833.2
Nebraska	1,586	1,410	1,522	4,518	685.4
Nevada	761	759	896	2,416	593.4
New Hampshire	1,093	1,077	1,092	3,262	747.6
New Jersey	6,193	5,844	6,476	18,513	661.2
New Mexico	1,206	1,257	1,310	3,773	792.5
New York	10,956	11,081	11,102	33,139	610.8
North Carolina	7,005	6,662	6,535	20,202	756.3
North Dakota	580	498	457	1,535	549.8
Ohio	9,802	8,743	8,855	27,400	745.4
Oklahoma	1,966	2,074	2,055	6,095	521.3
Oregon	1,433	1,563	1,636	4,632	582.7
Pennsylvania	13,218	12,433	12,172	37,823	907.0
Rhode Island	824	629	723	2,176	756.5
South Carolina	2,934	2,842	3,018	8,794	645.7
South Dakota	795	741	631	2,167	672.4
Tennessee	3,968	3,760	3,539	11,267	591.9
Texas	9,515	9,314	10,059	28,888	564.1
Utah	1,082	974	983	3,039	577.4
Vermont	387	397	392	1,176	518.3
Virginia	6,161	5,541	5,556	17,258	788.3
Washington	2,498	2,522	2,681	7,701	516.7
West Virginia	2,463	2,314	2,202	6,979	954.7
Wisconsin	4,246	4,365	4,789	13,400	670.7
Wyoming	692	686	556	1,934	1,172.1
Total	**189,116**	**181,374**	**187,205**	**557,695**	**664.2**

* *International Classification of Diseases, Ninth Revision, Clinical Modification* codes 416.0, 416.8, or 416.9 for any-listed diagnosis on Medicare hospital claims during 2000–2002.

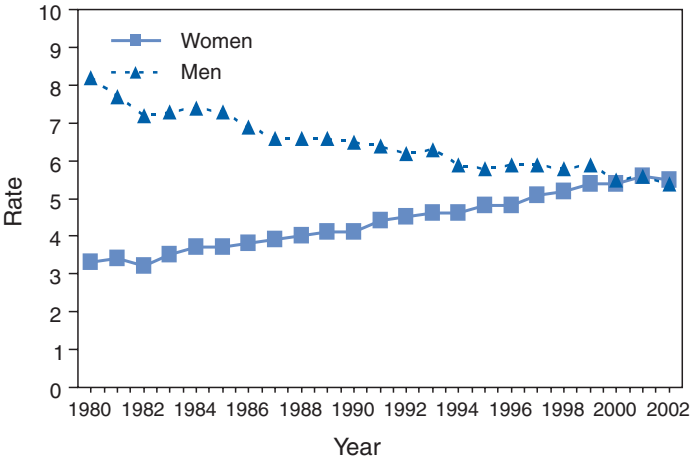
† Per 100,000 Medicare enrollees. Age-standardized to 2000 U.S. standard population aged ≥65 years.

TABLE 21. Comparison of disease reporting classifications of pulmonary hypertension with treatment-based classifications

International Classification of Diseases, Ninth Revision (ICD-9) classification*	*International Classification of Diseases, Tenth Revision* (ICD-10) classification†	Revised treatment-based clinical classification of pulmonary hypertension§
416.0 Primary pulmonary hypertension • Idiopathic pulmonary arteriosclerosis • Pulmonary hypertension (essential) (idiopathic) (primary)	I27.0 Primary pulmonary hypertension • Pulmonary (artery) hypertension (idiopathic) (primary)	1. Pulmonary arterial hypertension • 1.1. Idiopathic • 1.2. Familial • 1.3. Associated with –1.3.1. Collagen vascular disease –1.3.2. Congenital systemic-to-pulmonary shunts –1.3.3. Portal hypertension –1.3.4. HIV infection –1.3.5. Drugs and toxins –1.3.6. Other (thyroid disorders, glycogen storage disease, Gaucher disease, hereditary hemorrhagic telangiectasia, hemoglobinopathies, myeloproliferative disorders, and splenectomy) • 1.4. Associated with significant venous or capillary involvement: –1.4.1. Pulmonary veno-occlusive disease –1.4.2. Pulmonary capillary hemangiomatosis • 1.5. Persistent pulmonary hypertension of the newborn
416.8 Other chronic pulmonary heart diseases • Pulmonary hypertension, secondary	I27.8 Other specified pulmonary heart diseases	2. Pulmonary hypertension with left heart disease • 2.1. Left-sided atrial or ventricular heart disease • 2.2. Left-sided valvular heart disease 3. Pulmonary hypertension associated with lung diseases and/or hypoxia • 3.1. Chronic obstructive pulmonary disease • 3.2. Interstitial lung disease • 3.3. Sleep-disordered breathing • 3.4. Alveolar hypoventilation disorders • 3.5. Chronic exposure to high altitude • 3.6. Developmental abnormalities 4. Pulmonary hypertension attributed to chronic thrombotic and/or embolic disease • 4.1. Thromboembolic obstruction of proximal pulmonary arteries • 4.2. Thromboembolic obstruction of distal pulmonary arteries • 4.3. Nonthrombotic pulmonary embolism 5. Miscellaneous (Sarcoidosis, histiocytosis X, lymphangiomatosis, and compression of pulmonary vessels)
416.9 Chronic pulmonary heart disease, unspecified • Chronic cardiopulmonary disese • Cor pulmonale (chronic)	I27.9 Pulmonary heart disease, unspecified • Chronic cardiopulmonary disease • Cor pulmonale (chronic)	

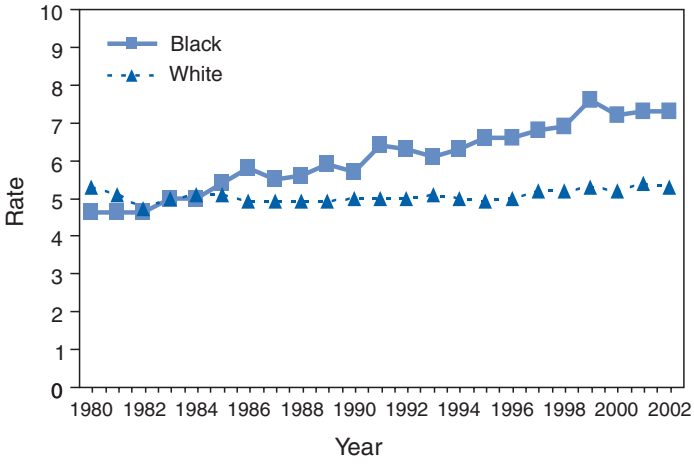
* World Health Organization. Manual of the international statistical classification of diseases, injuries, and causes of death, 9th revision, vol. 1. Geneva, Switzerland: World Health Organization; 1977.
† World Health Organization. International statistical classification of diseases and related health problems, 10th revision. Geneva, Switzerland: World Health Organization; 1992.
§ Simonneau G, Galie N, Rubin LJ, et al. Clinical classification of pulmonary hypertension. J Am Coll Cardiol 2004;43:5S–12S.

FIGURE 1. Age-standardized death rate* among decedents with pulmonary hypertension as any contributing cause of death, by sex and year — United States, 1980–2002



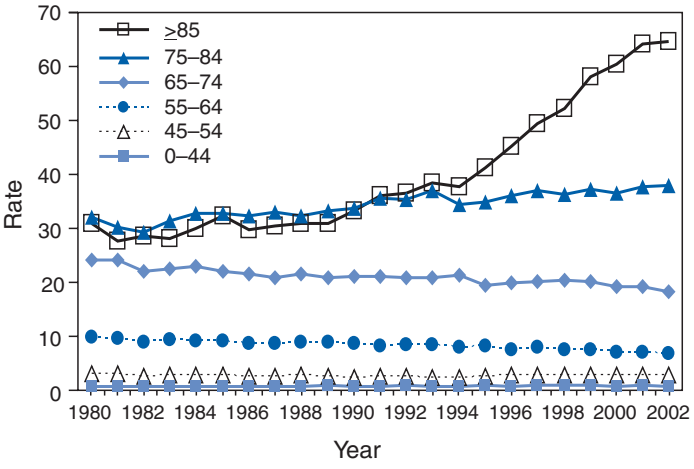
* Per 100,000 population. Age-standardized to the 2000 U.S. standard population.

FIGURE 2. Age-standardized death rate* among decedents with pulmonary hypertension as any contributing cause of death, by race† and year — United States, 1980–2002



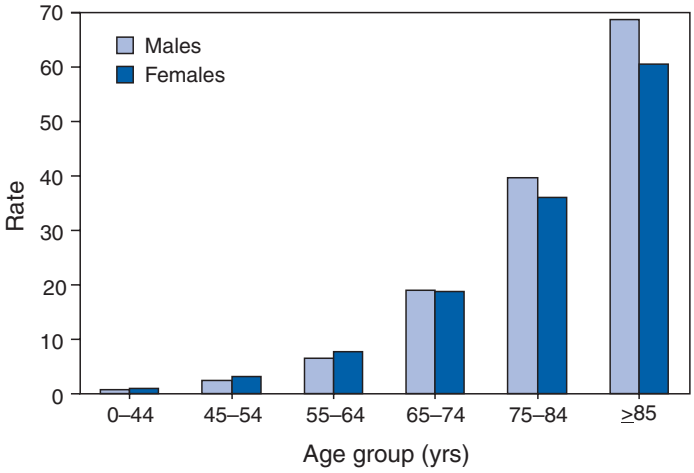
* Per 100,000 population. Age-standardized to the 2000 U.S. standard population.
† Data are presented only for white and black races because rates for other racial populations, when analyzed separately, were too small for meaningful analysis.

FIGURE 3. Age-specific death rate* among decedents with pulmonary hypertension as any contributing cause of death, by age group and year — United States, 1980–2002



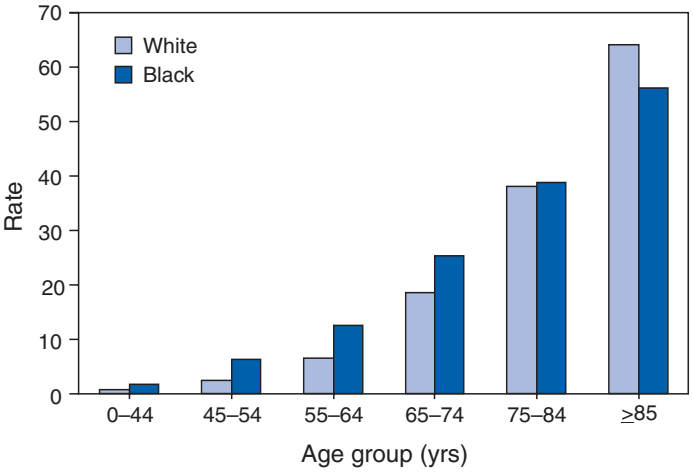
* Per 100,000 population.

FIGURE 4. Age-specific death rate* among decedents with pulmonary hypertension as any contributing cause of death, by sex and age group — United States, 2000–2002



* Per 100,000 population.

FIGURE 5. Age-specific death rate* among decedents with pulmonary hypertension as any contributing cause of death, by race† and age group — United States, 2000–2002



* Per 100,000 population.
† Data are presented only for white and black races because rates for other racial populations, when analyzed separately, were too small for meaningful analysis.

FIGURE 6. Age distribution among decedents with pulmonary hypertension as any contributing cause of death, by sex and age group — United States, 2000–2002

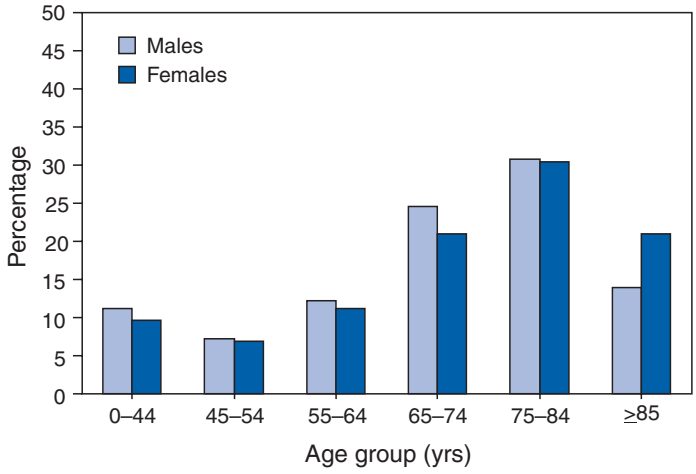
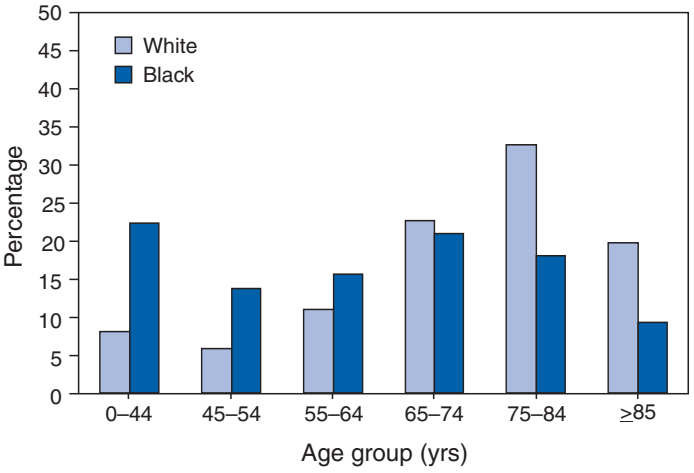
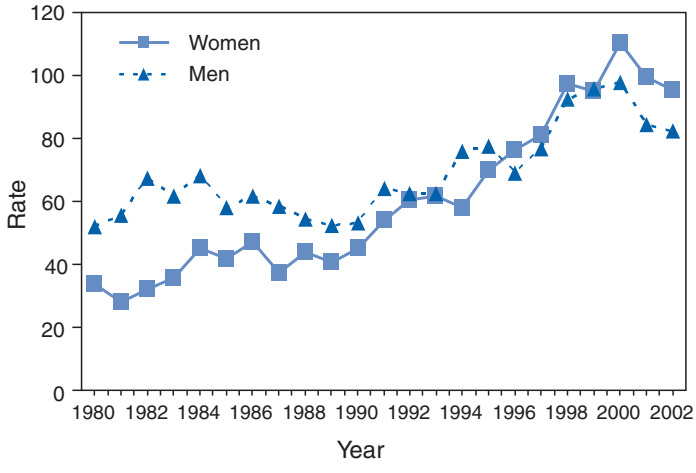


FIGURE 7. Age distribution among decedents with pulmonary hypertension as any contributing cause of death, by race* and age group — United States, 2000–2002



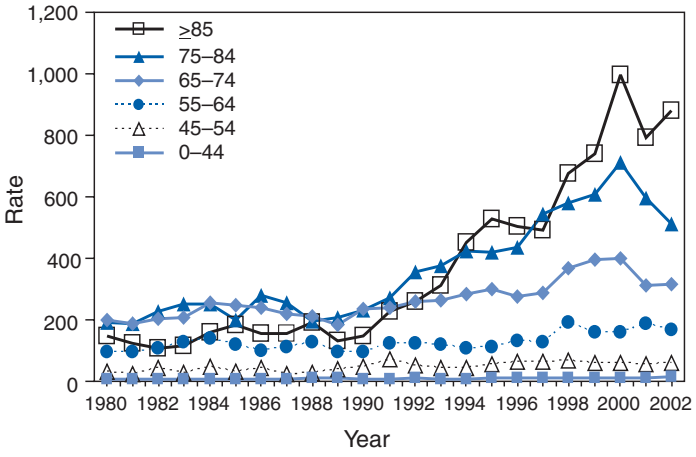
* Data are presented only for white and black races because percentages for other racial populations, when analyzed separately, were too small for meaningful analysis.

FIGURE 8. Age-standardized hospital rate* for persons with pulmonary hypertension as any-listed diagnosis, by sex and year — National Hospital Discharge Survey, United States, 1980–2002



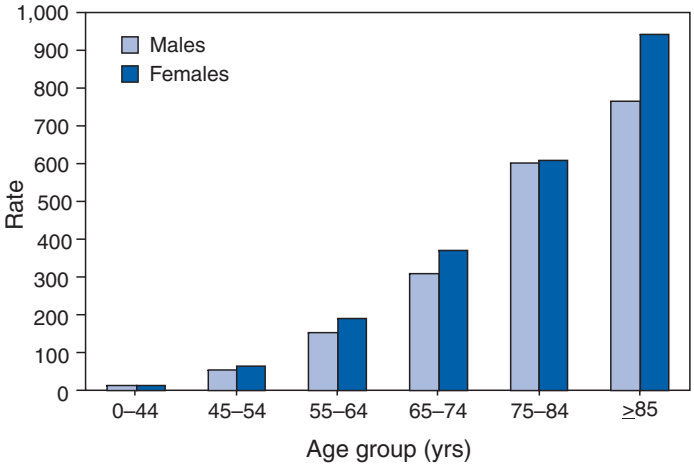
* Per 100,000 population. Age-standardized to the 2000 U.S. standard population.

FIGURE 9. Age-specific hospital rate* for persons with pulmonary hypertension as any-listed diagnosis, by year and age group — National Hospital Discharge Survey, United States, 1980–2002



* Per 100,000 population.

FIGURE 10. Age-specific hospital rate* for persons with pulmonary hypertension as any-listed diagnosis, by sex and age group — National Hospital Discharge Survey, United States, 2000–2002



* Per 100,000 population.

FIGURE 11. Age distribution among persons hospitalized with pulmonary hypertension, by sex and age group — National Hospital Discharge Survey, United States, 2000–2002

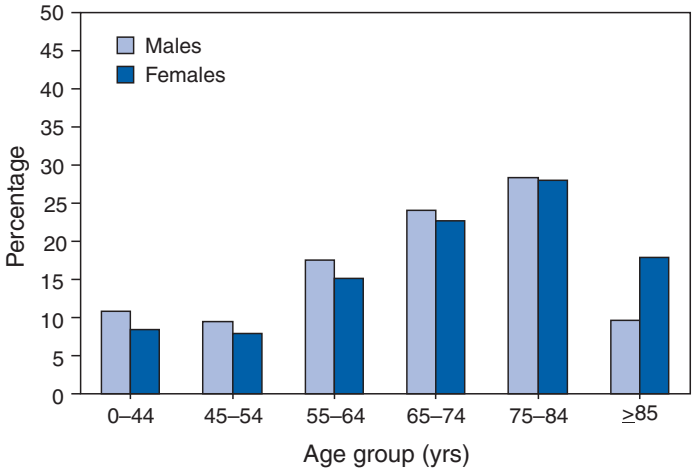
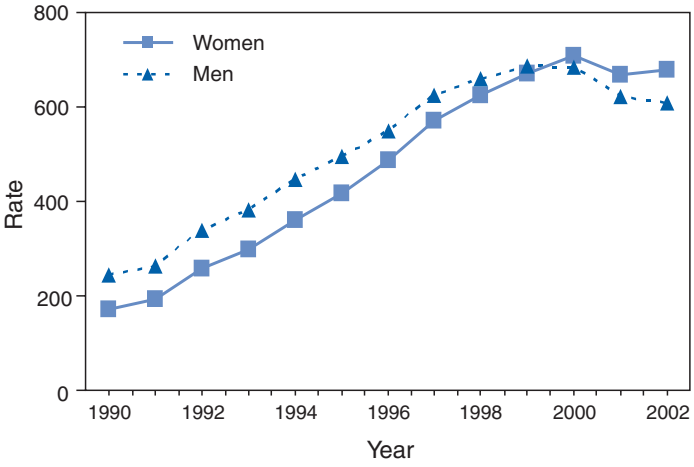
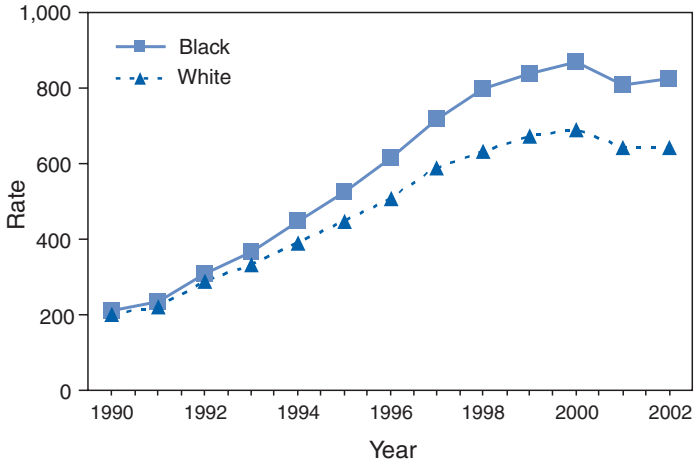


FIGURE 12. Age-standardized hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension as any-listed diagnosis, by sex and year — United States, 1990–2002



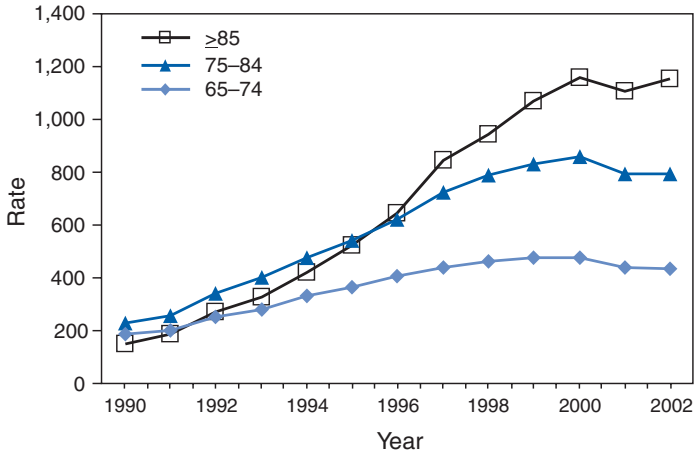
* Per 100,000 Medicare enrollees. Age-standardized to the 2000 U.S. standard population aged ≥65 years.

FIGURE 13. Age-standardized hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension as any-listed diagnosis, by race† and year — United States, 1990–2002



* Per 100,000 Medicare enrollees. Age-standardized to the 2000 U.S. standard population aged ≥65 years.
† Data are presented only for white and black races because rates for other racial populations, when analyzed separately, were too small for meaningful analysis.

FIGURE 14. Age-specific hospital rate* for Medicare enrollees aged ≥65 years with pulmonary hypertension as any-listed diagnosis, by age group and year — United States, 1990–2002



* Per 100,000 Medicare enrollees.

The *Morbidity and Mortality Weekly Report (MMWR)* Series is prepared by the Centers for Disease Control and Prevention (CDC) and is available free of charge in electronic format and on a paid subscription basis for paper copy. To receive an electronic copy each week, send an e-mail message to *listserv@listserv.cdc.gov*. The body content should read *SUBscribe mmwr-toc*. Electronic copy also is available from CDC's World-Wide Web server at *http://www.cdc.gov/mmwr* or from CDC's file transfer protocol server at *ftp://ftp.cdc.gov/pub/publications/mmwr*. To subscribe for paper copy, contact Superintendent of Documents, U.S. Government Printing Office, Washington, DC 20402; telephone 202-512-1800.

Data in the weekly *MMWR* are provisional, based on weekly reports to CDC by state health departments. The reporting week concludes at close of business on Friday; compiled data on a national basis are officially released to the public on the following Friday. Address inquiries about the *MMWR* Series, including material to be considered for publication, to Editor, *MMWR* Series, Mailstop K-95, CDC, 1600 Clifton Rd., N.E., Atlanta, GA 30333; telephone 888-232-3228.

All *MMWR* references are available on the Internet at http://www.cdc.gov/mmwr. Use the search function to find specific articles.

Use of trade names and commercial sources is for identification only and does not imply endorsement by the U.S. Department of Health and Human Services.

References to non-CDC sites on the Internet are provided as a service to *MMWR* readers and do not constitute or imply endorsement of these organizations or their programs by CDC or the U.S. Department of Health and Human Services. CDC is not responsible for the content of these sites. URL addresses listed in *MMWR* were current as of the date of publication.

☆U.S. Government Printing Office: 2006-523-142/00123 Region IV ISSN: 1546-0738