MMWR article describes two of nine varicella deaths reported in the United States in 2002

This article appears in today’s (June 13, 2003) issue of Morbidity and Mortality Weekly Report (MMWR), a publication of the Centers for Disease Control and Prevention (CDC). Titled “Varicella-Related Deaths—United States, 2002,” the article states that nine people were reported to have died from complications of varicella in 2002. The article also presents case reports on a 37-year-old man and 9-year-old-girl who died after exposure to unvaccinated children with varicella disease. Residents of Kansas and Illinois, respectively, the man and girl had been healthy before exposure to the disease.

Varicella has been a vaccine-preventable disease in the United States since 1995, when varicella vaccine was licensed. The “Recommended Childhood and Adolescent Immunization Schedule—United States, 2003” specifies one dose of varicella vaccine for children age 12 to 18 months and catch-up immunization for susceptible children and adolescents age 2 through 18 years.

The article illustrates the consequences of the failure to establish immunity in a population. By early 2002, 23 states and the District of Columbia had implemented varicella vaccination requirements for child care or school entry. Kansas and Illinois were not among them.

Varicella-related Deaths—United States, 2002

Varicella is a vaccine-preventable disease that can be fatal. During 2002, state health departments notified CDC about nine fatal cases of varicella in adults and children. This report summarizes clinical data for one adult and one child, reported from Kansas and Illinois, respectively. Both patients were susceptible, unvaccinated, and exposed to unvaccinated children with varicella. These deaths highlight the importance of implementing strategies recommended for varicella disease prevention, including child care and school vaccination requirements, and underscore the need for improving varicella death surveillance.

Case Reports

Case 1. On January 19, 2002, an immunocompetent man aged 37 years reported to an emergency department (ED) with acute cough and shortness of breath preceded by a 3-day history of skin rash and a 4-day history of fever. He was exposed to his unvaccinated daughter aged 9 years, who had varicella disease (rash onset: January 3). The patient’s other daughter aged 5 years (also unvaccinated) had rash onset 2 days after her father’s. Before the patient’s admission, neither he nor his children had been examined by a health-care provider for varicella-related signs or symptoms. The patient had no history of varicella and was unvaccinated. His medical history included current smoking.

On initial examination, the patient had numerous skin lesions consistent with varicella and diffuse inspiratory crackles. Chest radiography showed a five-lobe interstitial infiltrate with slight nodularity, suggestive of varicella pneumonia. Intravenous acyclovir, broad-spectrum antibiotic therapy, and oxygen were initiated. The patient was admitted to the intensive care unit. Overnight, his respiratory difficulty increased, and he required intubation.

During hospitalization, the patient had complications including recurrent pneumothoraces, cardiopulmonary arrest, anoxic encephalopathy, bacteremia (methicillin-resistant coagulase negative staphylococcus), left upper extremity deep venous thrombosis, and coma. He died on March 9.

Laboratory tests of nasopharyngeal specimens were negative for influenza A and B antigens. An autopsy was not performed.

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Case 2. On January 14, a girl aged 9 years was taken to an ED with a 3-day history of classic varicella rash, a 2-3 day history of inability to bear weight on the left foot and leg, and a history of fever of unspecified duration. The patient had no history of varicella and was unvaccinated. Her history was negative for traumatic injury.

On initial examination, the patient had fever (101 degrees F [38.3 degrees C]), a generalized rash with crusted lesions, and mild swelling, induration, and warmth over the left calf, ankle, and foot. The patient was admitted with diagnoses of varicella, possible sepsis, and possible left lower extremity cellulitis. Intravenous nafcillin was ordered. Approximately 12 hours after initial evaluation, purple discoloration surrounding the patient’s varicella lesions was noted. Subsequently, the patient had respiratory distress and, despite intubation, cardiac arrest ensued. The patient died approximately 16 hours after initial assessment. Premortem blood cultures yielded beta-hemolytic Streptococcus pyogenes group A.

Autopsy revealed multiple scabbed lesions consistent with varicella, intravascular thrombi, increased fluid in the pericardial sac, bilateral pulmonary edema and congestion, hepatic and splenic congestion, and a left lower extremity calf circumference 2 cm greater than that of the right calf. No evidence of a saddle pulmonary thromboembolus was noted.

The patient had been exposed in after-school child care to an unvaccinated child aged 7 years with varicella (rash onset: December 20, 2001) and in school to two unvaccinated children with varicella (rash onset: December 21).

Editorial Note:
The cases described in this report demonstrate the potential seriousness of varicella disease. With the licensure of a safe and effective varicella vaccine in 1995, varicella became a vaccine-preventable disease. Prevention of varicella-related deaths through vaccination should be a public health priority. During 1990-1994, before implementation of the varicella vaccination program, an estimated 4 million cases, 11,000 hospitalizations, and 100 deaths were attributed to varicella disease each year in the United States. As with the patients described in this report, the majority of persons who died of varicella during 1990-1994 were previously healthy.

In 1995 and 1996, respectively, the American Academy of Pediatrics and the Advisory Committee on Immunization Practices (ACIP) recommended that all children aged 12-18 months be vaccinated routinely and that all susceptible children be vaccinated by age 13 years. In addition, ACIP recommended vaccination for susceptible persons who have close contact with persons at high risk for serious complications (e.g., health-care workers and family contacts of immunocompromised persons). In 1999, ACIP expanded its recommendations to promote varicella vaccination for susceptible persons in the following high-risk groups: 1) persons who live or work in environments in which transmission of varicella is likely (e.g., teachers of young children, child care employees, and residents and staff members in institutional settings), 2) persons who live and work in environments in which transmission can occur (e.g., college students, inmates and staff members of correctional institutions, and military personnel), 3) nonpregnant women of child-bearing age, 4) adolescents and adults living in households with children, and 5) international travelers. ACIP also recommended postexposure vaccination for susceptible persons.

Varicella disease was not nationally reportable in 1995 when the vaccine was introduced. As a result, no national data were available to monitor the impact of the vaccination program. In 1995, CDC, in collaboration with state and local health departments, instituted active surveillance in three communities. In 2000, disease and hospitalizations in these areas declined approximately 80% compared with 1995. Herd immunity probably contributed to these trends. This hypothesis is supported by the observation of declines in all age groups, including children aged less than 1 year, who are ineligible for vaccination, and persons aged over 20 years, who are not highly vaccinated. This hypothesis is further supported by declines occurring at vaccine coverage (continued on next page)
levels of 74%-84% among children aged 19-35 months. Disease rates are expected to decline further with improved coverage.

In 2001, state-specific varicella vaccination coverage in the United States among children aged 19-35 months ranged from 53% to 90%. Vaccination coverage among children aged more than 35 months is unknown. If each state implements child care and school entry vaccination requirements as recommended by ACIP in 1999, high nationwide coverage will be achieved. The recommendations specify that children entering child care facilities and elementary schools in every state should be required either to have received varicella vaccine or to demonstrate other evidence of varicella immunity. By December 2002, a total of 34 (67%) states had implemented child care and/or school laws (CDC, unpublished data, 2002). Requirements differ among states, applying to children at one or more levels of education (i.e., kindergarten, elementary school, middle school, and high school). When the two deaths described in this report occurred, neither Kansas nor Illinois had implemented child care or school entry vaccination requirements.

Active surveillance data demonstrate morbidity reduction since initiation of the varicella vaccination program, but national disease data are unavailable. In 1999, in initiating national varicella surveillance, the Council of State and Territorial Epidemiologists mandated reporting of varicella-related deaths to CDC’s National Immunization Program (NIP). To date, substantial underreporting of varicella-related deaths to NIP continues to occur, and the use of limited mortality data in assessing the impact of the varicella vaccination program remains difficult. According to National Center for Health Statistics (NCHS) data for 2000, varicella was listed in death certificates as the primary cause of death for 44 deaths reported by 23 states and the District of Columbia; however, only nine (20%) varicella-related deaths were reported to NIP by seven states (CDC, unpublished data, 2002). Reporting to NIP complements NCHS data. Data submitted to NIP include detailed case information that allows examination of each patient’s risk factors for morbidity and mortality.

Through adherence to current varicella vaccination recommendations, further reduction of varicella-related morbidity and mortality can be achieved and sustained in the United States (CDC, unpublished data, 2002). More widespread implementation of child care and school vaccination requirements (including those for middle and high school) will ensure that children who are not infected during childhood because of decreasing varicella virus circulation will be protected by vaccination before reaching adulthood, when their risk for severe disease and complications is increased. When susceptible persons are exposed, they should be vaccinated. When disease severity necessitates hospitalization or results in death, laboratory confirmation of disease should be considered. When patients die from varicella or associated complications, a varicella-related death investigation worksheet, available through state health departments, should be completed. State personnel should fax or mail investigation worksheets (without personal identifiers) to NIP, fax (404) 639-8665. For reporting assistance, state health departments should contact NIP’s Viral Vaccine-Preventable Disease Branch, telephone (404) 639-8230.