Mortality Due to Farmer’s Lung in Finland*

Jouko Kokkarinen, M.D.; Hannu Tukiainen M.D.; and Erkki O. Terho, M.D.

Fatal cases of farmer’s lung (FL) are rare. We found, based on death certificates, 13 cases of FL with a fatal outcome in Finland between 1980 and 1990. Compared with incidence data from the years 1980 to 1982, the mortality was estimated as 0.7 percent. On average, death occurred 8 years after the diagnosis of FL. One patient died acutely after a heavy mold exposure. The other patients had chronic disease. Among them the immediate cause of death was pneumonia in seven patients, respiratory insufficiency in four, and pulmonary fibrosis in one patient. The majority of these patients with a fatal outcome had suffered from symptoms of FL for more than 1 year before the diagnosis was established and fibrotic changes were already visible in the chest radiograph at the time of the diagnosis.

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Acute farmer’s lung (FL) usually has a good prognosis. However, in some patients the disease leads to a chronic phase with pulmonary fibrosis that may lead to death. A fatal case of acute FL has also been reported. In long-term follow-up studies, the mortality associated with FL has ranged from 0 to 17 percent.

To assess the mortality of FL, we have reviewed death certificates and clinical data of patients who had hypersensitivity pneumonitis as the underlying cause of death in Finland between the years of 1980 and 1990.

MATERIALS AND METHODS

In Finland, the death certificates are sent from the doctor signing the document to local and provincial medico-legal officers. After checking, the causes of death are registered in the Central Statistical Office. Death certificates were coded according to a Finnish edition, 1969, of WHO International Classification of Diseases, 1967, eighth revision up to 1986 and thereafter according to a Finnish edition, 1987, of WHO International Classification of Diseases, 1977, ninth revision. Patients with hypersensitivity pneumonitis were searched for with the ICD code 516 up to 1986 and with the ICD code 495 thereafter. The code 516 in the Finnish Classification of Diseases, 1969, included pneumoconioses and lung diseases due to nonorganic (excluding silicosis and asbestosis) and organic (including hypersensitivity pneumonitis) agents and fumes. The code 495 in the Finnish Classification of Diseases, 1987, included farmer’s lung and other forms of hypersensitivity pneumonitis. The search with these codes revealed 22 death cases from 1980 to 1990. Permission to review the patient files of these patients was obtained from the Ministry of Social Affairs and Health.

For the diagnosis of FL each case had to fulfill the clinical criteria of FL. These criteria consisted of three main and six additional criteria. The main criteria were (1) exposure to offending antigens, revealed by history, by aeroallergologic investigations of the environment, or by the measurements of antigen-specific immunoglobulin G antibodies; (2) symptoms compatible with FL; and (3) lung infiltrations on chest radiographs compatible with FL. The additional criteria were (1) basal crepitant rales audible on auscultation of the lungs, (2) impairment of pulmonary diffusing capacity, (3) arterial oxygen tension (or saturation) decreased either at rest or during exercise, (4) restrictive ventilatory defect in the spirometry, (5) histologic changes compatible with FL in a biopsy specimen from the lung, and (6) positive provocation test. The diagnosis was considered confirmed if the patient fulfilled all the main criteria and at least two of the additional criteria and if all other diseases with similar symptoms had been ruled out. If the criteria were otherwise fulfilled, but the chest radiograph was normal, the diagnosis was considered confirmed if a lung biopsy was compatible with FL.

The FL was considered as an underlying cause of death when the disease had progressed to advanced pulmonary fibrosis and the immediate cause of death was respiratory, eg, respiratory insufficiency or pneumonia. Also, one case where the patient died acutely after a massive mold exposure which was confirmed as FL by an autopsy was included.

Four of the 22 patients had not worked on a farm. The causes of death in these four patients were hypersensitivity pneumonitis in a florist with a moldy basement floor in her workplace, sarcoidosis, COPD, and unspecified pneumoconiosis each in one patient. In addition, there was one exfarmer, who had stopped farming years before the onset of the ultimately fatal disease. In this patient, open lung biopsy specimen was indicative of hypersensitivity pneumonitis, but the exact cause of the disease remained obscure.

Thirteen of the 17 patients with farming history were considered to have FL as the underlying cause of death. Out of the remaining four patients, two had FL but its contribution to the death remained uncertain. One died suddenly at his home 2 months after the diagnosis of FL for an unknown reason. The other died 8 years after the diagnosis of FL due to myocardial infarction. One patient was regarded to have pulmonary fibrosis of unknown cause, one died at the age of 81 from pulmonary fibrosis. In the latter case, there was a histologic suspicion of hypersensitivity pneumonitis in the autopsy, but there were no clinical data of possible exposure to causative factors. Thus, 13 cases of death

*From the Department of Pulmonary Diseases (Drs. Kokkarinen, Tukiainen, and Terho), Kuopio University Hospital, Kuopio, Finland; and the Department of Clinical Allergology (Dr. Terho), University of Turku, Turku, Finland.

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Reprint requests: Dr. Kokkarinen, Department of Pulmonary Diseases, Kuopio University Hospital, SF 70210 Kuopio, Finland
remained for final analysis.

Mortality of FL was calculated on the basis of the incidence data of the disease during 1980 to 1982. During these years, new cases of clinically confirmed FL based on criteria presented by Terho were registered in all special hospitals for pulmonary diseases and in departments of pulmonary diseases in other hospitals in Finland. During that time, 512 new cases of FL were recorded, 186 in 1980, 153 in 1981, and 173 in 1982. At the time of the diagnosis of FL, the mean age of the patients was 47 years. Two-thirds (67 percent) of the patients were women.

RESULTS

The clinical data of the 13 patients with FL as the underlying cause of death are presented in Table 1. All patients fulfilled the diagnostic criteria of FL presented by Terho and all had chest radiograph changes compatible with FL. The mean age of the patients was 52 years at the time of the diagnosis and 54 percent of them were women. Data of smoking habits were available for eight patients: all were nonsmokers. Before the diagnosis, 8 of 13 patients had suffered from symptoms of FL for more than 1 year. All the patients were full-time or part-time dairy farmers. Mold exposure in the cowhouse resulted usually from moldy hay or bedding.

Pulmonary function data were available only for some of the patients and were usually expressed as a percentage of the predicted values (Table 1). Because the fatal cases were scattered throughout the country, different pulmonary function equipment and reference values were used, details of which were usually not available in the patient files. When measured, pulmonary diffusing capacity was impaired. Reduced forced vital capacity (FVC) (<80% of pred) was seen in six of ten patients.

At the time of the diagnosis of FL, 11 patients were treated with corticosteroids, usually with oral prednisolone. The patient with an acute fatal outcome of the disease had treatment with intravenous methylprednisolone. Data of the duration of corticosteroid treatment were available for eight patients: the duration varied from 6 weeks to 7 months.

Of 12 patients with follow-up data, 6 returned to work in the cowhouse after the diagnosis of FL. Chest radiograph normalized in one of these patients during the first 6 months after the diagnosis, and fibrotic changes remained in three patients. For two patients, data on radiographic recovery during the first year were not available. All these six patients later experienced recurrent attacks of the disease. The other six patients did not return to regular work in the cowhouse, although two of them still worked there occasionally. Radiographically, fibrotic changes persisted in five of these patients. For one patient, data concerning recovery of chest radiograph during the first year was unavailable.

At the time of the diagnosis of FL, four patients had arterial hypertension and one patient suffered from diabetes. Later diabetes was diagnosed in three, arterial hypertension in one, coronary artery disease in one, and asthma and pulmonary tuberculosis in one patient. *Cor pulmonale* was diagnosed in eight patients. Four patients were on supplemental oxygen during the terminal phase of the illness.

Death occurred more than 5 years after the diagnosis of FL in 10 of 13 patients (Table 1). One patient died acutely after 1 day following a heavy mold exposure (sacking moldy grain). In the other 12 patients, the immediate cause of death was pneumonia in 7, respiratory insufficiency in 4, and pneumothorax in 1 patient.

Autopsy was made in six patients. The patient, who

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<th>Patient No.</th>
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<th>Duration of Symptoms, mo</th>
<th>FVC* (% pred)</th>
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*At the time of the diagnosis of FL.

†Symptoms had lasted for years, but no accurate data was available.

Table 1—Clinical Data of Fatal FL Cases
died acutely after a heavy mold exposure had mononuclear cell infiltration in the lung and also numerous granulomas with epithelioid cells and giant cells. Fibrosis indicative of a chronic disease was seen. There were also patchy areas with abundant granulocytes in the lung compatible with pneumonia. In the other five patients, autopsy showed advanced pulmonary fibrosis. In three patients, there were mononuclear cell infiltration or scanty giant cells but no clear granulomas were found. In two patients pneumonia was evident. One patient had pulmonary tuberculosis and pneumothorax.

Compared with the incidence data of FL during the period of 1980 to 1982, the mortality was calculated to be 0.7 percent.

DISCUSSION

We calculated the mortality based on data about the incidence of FL in the entire country from the years 1980 to 1982. These incidence data are representative, for in that nationwide study the same diagnostic criteria were used as in our study. Furthermore, the time at the beginning of 80s was close to the period when the diagnoses of FL were made in our series. We included only cases in which FL for a certainty was considered as an underlying cause of death.

We found 13 fatal cases of FL between 1980 and 1990 in Finland that resulted in a mortality of about 1 percent. In two Finnish studies with a mean follow-up of 5 years and one Canadian follow-up study no fatal cases of FL were reported. Three long-term follow-up studies in the 1960s and 1970s reported mortality of 9 to 17 percent associated with FL.

Emanuel and coworkers reported four fatal cases in a series of 24 patients. All of them had developed progressive pulmonary fibrosis as a consequence of FL. Barbee and coworkers reported five fatal cases in a series of 50 patients with a mean of 6 years of follow-up. All these patients had extensive lung disease. Braun and coworkers followed up 141 patients with FL. During a mean of follow-up of 14.8 years, 9 percent died because of FL. Death occurred, on the average, 17 years after the diagnosis of FL.

Cuthbert and Gordon reported 2 deaths in a follow-up study of 31 FL patients, but both patients died from myocardial infarction. Seal and coworkers described five cases with chronic FL as the main cause of death.

One of our patients died acutely after a heavy mold exposure. In the autopsy, there was evidence for fibrosis in addition to acute changes of FL, indicating long-standing disease. Barrocliff and Arblander have reported an acute fatal case of FL with the first exposure to moldy hay occurring only weeks before the death.

All the other patients in our study had chronic FL. Death was attributed to respiratory insufficiency or pneumonia, and in one patient to pneumothorax. Two patients had pneumothoraces during the course of the illness. Emanuel and coworkers also reported recurrent pneumothoraces in an ultimately fatal case of FL.

Eight of our 13 fatal cases had suffered from symptoms of FL for more than 1 year before the diagnosis was made. This is considerably longer time than in our other series of 101 patients with FL, where 37 percent of patients had symptoms of FL more than 3 months before the diagnosis. Also in bird fancier’s lung, a long symptomatic time before the diagnosis has been considered to be a major factor leading to the development of chronic disease.

The mainstay in the treatment of FL is the strict avoidance of the offending antigens. Nonetheless, the majority of the patients can return to their previous work after full recovery from the acute episode. Exposure to the offending antigens is reduced by changes in farming techniques and by the use of personal dust respirators. The majority of our fatal cases had at the time of the diagnosis permanent fibrotic changes visible in chest radiograph indicating chronic FL, and six patients who returned to their previous work had recurrent attacks of the disease. When also minimal changes are included, chronic fibrotic changes have been reported in 28 percent of patients at the time of the diagnosis of FL.

In conclusion, based on death certificates and incidence data of FL, we found about 1 percent mortality attributable to FL. Death occurred an average of 8 years after the diagnosis. Majority of the patients had a long symptomatic period before FL was diagnosed and had fibrotic changes in chest radiograph already at the time of the diagnosis of FL.

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