

INTERSTITIAL FIBROSIS IN COAL WORKERS— EXPERIENCE IN WALES AND WEST VIRGINIA

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Abstract—Forty-five Welsh coal-miners with interstitial pulmonary fibrosis have been studied in detail. They differ from other members of the population with this disease by having significantly longer survival. Mean duration of illness was 11.4 ± 5 years for those who had died compared with a mean maximum survival of 4.5 years quoted in published series of pulmonary fibrosis of non-occupational origin. The disease is not unique to South Wales coal-miners. Autopsy studies from two populations of United States miners suggest a similar prevalence. Pathological features are the same in both groups.

INTRODUCTION

INTERSTITIAL PULMONARY FIBROSIS (I.P.F.) occurs in coal-workers for reasons unknown. In this respect the disease is similar to that seen in the general population. However, in South Wales, increasing recognition of this condition both in life and at post-mortem has led to the hypothesis that there may be an association between inhalation of coal dust and the development of I.P.F. Over the past eight years, coal-miners with I.P.F. attending the Medical Research Council Pneumoconiosis Unit (MRCPU) in Cardiff have been studied. Clinical, pathological and radiological features of 45 men have been examined in detail to characterise the disease in this group. In order to determine whether Welsh coal-miners are unusual, we have compared our findings with two populations of coal-miners employed in the U.S.A.

SUBJECTS AND METHODS

All new patients attending the MRCPU from May 1977 were scrutinized. Those considered to have interstitial pulmonary fibrosis (IPF) were selected for this study. Many of these patients had been referred by the Cardiff Pneumoconiosis Medical Panel (PMP) which also contributed further patients collected over the previous years. Thus they were a highly selected group unsuitable for estimations of incidence. It is likely that more cases exist than have come to our attention.

There is no strict clinical definition of IPF. We used evidence of gradually progressive breathlessness associated with finger clubbing and/or showers of fine fixed inspiratory crackles at the lung bases, along with predominantly irregular shadows on the chest radiograph and physiological evidence of parenchymal damage. The clinical

data were extracted from notes kept by the MRCPU and the PMP. Occupational histories were obtained in every case, generally in considerable detail. Any suggestion of asbestos exposure was carefully noted. All but one admitted cigarette smoking, thus verification was not considered essential. A search was made for any features suggestive of extrinsic allergic alveolitis with particular note taken of hobbies such as pigeon racing. Similarly, other possible causes of pulmonary fibrosis were sought. Thus the men were considered to have IFP of unknown origin.

Lung function tests were all performed at the MRCPU using a standardized technique and figures were compared with reference values (COTES, 1979). Forty-three patients had estimates of lung volumes and gas transfer factor in addition to spirometry. Measurements were made throughout the course of the disease, but not necessarily in a systematic manner. The results obtained closest to time of death are reported. No attempt was made to correlate functional changes with pathological findings.

All available chest radiographs (CXR) for each man were examined by three readers experienced in the use of the ILO classification (INTERNATIONAL LABOUR OFFICE, 1980). Where a series of films existed, the first film to show predominantly irregular opacities was used in conjunction with the clinical findings to determine onset of the disease.

Post mortem material was available on 19 of the 23 men dead at the time of analysis. The lungs were assessed semi-quantitatively using an agreed protocol for type and severity of pneumoconiosis and IPF.

United States Autopsy Populations

Two populations of autopsied U.S. coal miners were evaluated for the prevalence of interstitial lung disease at death.

The National Coal Workers' Autopsy Study (NCWAS) Population

The National Coal Workers' Autopsy Study (NCWAS) and Disaster Plan was initiated under section 203 (d) of the Federal Coal Mine Health and Safety Act in 1969 and is now conducted under the amendment to the Act known as the Federal Mine Safety and Health Act of 1977. This is a voluntary programme administered by the National Institute for Occupational Safety and Health (NIOSH). Participating pathologists submit the cases to NIOSH and include an autopsy report with a minimum of three blocks of lung tissue and corresponding slides. Demographic information, occupational histories and smoking histories are available for almost all cases. Since its inception in 1972 to the end of 1980, 3,341 autopsies have been accepted from 27 states. The average age of the population is 62 ± 12 years with an average work period of 26 ± 14 years underground. Seventy-five per cent had been cigarette smokers.

The NCWAS population is similar with regards to geographic distribution, smoking status, and specific mining job categories to active miners in the NIOSH National Coal Workers' study. Sections of lung tissue were reviewed in a 12% sample of all cases received (disaster and routine) in order to obtain an estimate of the prevalence of IPF and pneumoconiosis (CWP). CWP lesions were categorised according to the criteria developed by the Pneumoconiosis Committee of the College of American Pathologists in conjunction with NIOSH (KLEINERMAN, *et al.*, 1979). In this sample interstitial fibrosis was determined as either present or absent.

Southern West Virginia Autopsy Population

Between 1959 and 1973, 694 underground coal miners were autopsied by Dr. W. Laqueur and colleagues at the Beckley Appalachian Regional Hospital in southern West Virginia. The study was conducted under a Public Health Service grant. Consent for autopsy was requested from all miners who died in the hospital during this period and was obtained in approximately 60%. Whole left lung sections were prepared (GOUGH and WENTWORTH, 1949) following fixation by intratracheal perfusion with 10% buffered formalin at a controlled pressure of 30 cm of water. All cases were reviewed grossly for evidence of honeycombing and for evidence of severe generalised emphysema associated with diffuse black pigmentation. Histological sections from a 17% sample ($n = 118$) were evaluated using the same protocol as the S. Wales group.

RESULTS

Clinical Data (MRCPU patients)

At the time of the study, 23 patients had died and 22 were still alive. For the group as a whole, mean survival was 9.7 ± 5 years (range 1–23 years). Mean survival for those who had died was 11.4 ± 4.5 (range 4–23 years) and for those still alive 7.9 ± 5 (range 1–21 years). Mean age at diagnosis was 55.5 ± 7 years (range 41–70 years).

Clubbing was noted in 31 (69%) and crackles in 37 (82%). Twenty-seven (60%) had both clubbing and crackles and 4 had neither.

All of the patients were male and all had been exposed to coal dust at underground coal mines in the Welsh valleys. Mean time spent working underground was 33.7 ± 12.4 years (range 6–51 years). Fifteen had worked purely on the coal face. The remainder had had a variety of jobs: seven were predominantly repairers but many were notable for the variety of different jobs undertaken. All but one man admitted regular cigarette smoking at some time. Fifteen were noted to be ex-smokers with 29 continuing to smoke throughout life.

The most consistent physiological finding was reduced gas transfer in 42 of the 43 cases studied.

Irregular opacities were the predominant radiological abnormality. Forty-four men achieved category 2 for overall profusion of small opacities as judged by the 1980 ILO Classification of chest radiographs.

Table 1 shows estimated year of onset for all patients. There is no evidence to suggest that we are recognising more cases now than previously.

TABLE 1. ESTIMATED YEAR OF ONSET FOR ALL PATIENTS

Estimated year of onset	Number
1955–60	8
1961–65	5
1966–70	10
1971–75	6
1976–80	10
1981–85	6

Table 2 shows length of survival both for patients still alive and for those who have died. Considering the average age at onset, a 10 year survival of 65% appears favourable.

The most common pathological finding in the MRCPU patients was diffuse interstitial fibrosis with and without coal dust pigmentation (Table 3) but numbers are too small to allow correlations between pathological diagnosis and outlook. Of those in whom pigmented fibrosis was noted, all but one had continuing exposure to coal dust underground after onset of their illness. The remaining man may also have had further exposure to coal dust in his job attending to school furnaces.

National Coal Workers' Autopsy Study

The overall prevalence of any form of CWP was found to be 64.25%; 55.6% showed soft dust associated focal macules with emphysema, 28% showed nodular lesions, 11.6% showed typical silicotic lesions and 6.2% had progressive massive fibrosis (PMF). Interstitial fibrosis was found in 15.9% of the population. Many miners showed evidence of two or more forms of disease.

South West Virginia Autopsy Population

Gross examination revealed 2.3% with honeycomb features and a further 7.6% had generalized air space enlargement with diffuse black pigmentation. Microscopic survey of a 17% sample using a common protocol showed 33% had interstitial fibrosis. Of these, 17.8% had moderate or severe forms of the disease. In this group, coal dust pigmentation of the fibrosis was present in over half, but it was the predominant

TABLE 2. PERCENTAGE OF PATIENTS ALIVE AT 5, 10 AND 15 YEARS AFTER ONSET

	Men who have died <i>n</i> = 23	Men still alive <i>n</i> = 22	Total <i>n</i> = 45
Survived for:			
5 years	96	73	84
10 years	65	32	49
15 years	22	14	18

TABLE 3. MRCPU PATIENTS—RELATIONSHIP BETWEEN SURVIVAL AND POST-MORTEM FINDINGS

Pathology	Survival in Years			
	0-5	6-10	11-15	15+
Slight to moderate IPF without coal pigment	0	1	1	1
with coal pigment	0	1	0	0
Diffuse IPF without coal pigment	0	3	1	1
with coal pigment	1	2	1	0
Cystic fibrosis/honeycomb without coal pigment	0	2	0	1
with coal pigment	0	3	0	0

feature in only 10%. In contrast, 47% had predominantly unpigmented fibrosis. Pigmented lesions were associated with emphysema and with more severe forms of pneumoconiosis particularly nodular silicotic lesions. They were not infiltrated with chronic inflammatory cells. The unpigmented lesions were not associated with emphysema and usually showed diffuse infiltration with chronic inflammatory cells and lymphoid aggregates.

DISCUSSION

There is no doubt that interstitial pulmonary fibrosis occurs in coal workers, but it is unknown whether this is related to coal dust inhalation. IPF in the general population is described as a condition recognizable by the pattern of progressive breathlessness occurring predominantly in the 4th and 5th decades, often accompanied by finger clubbing and crackles in the chest with physiological evidence of a restrictive lung defect with low gas transfer (Table 4) and a chest radiograph showing irregular opacities. All series, including this one, have similar characteristics, except for outcome (Table 5). The prognosis for these coal miners appears significantly better.

It is not clear from the survey why this should be so. Coal workers may possibly develop a different form of pulmonary fibrosis from IPF of unknown cause, but previous series of IPF, which included cases of pulmonary fibrosis produced by known aetiological factors such as collagen vascular disease (TURNER WARWICK, *et al.*, 1980; TUKIAINEN, *et al.*, 1983), showed no differences in outcome between the two groups.

Bias in the collection of cases may well have been responsible for differences found. However, all the series reported were biased by including only those cases who have come to medical attention. Coal miners, who have routine chest radiography are more likely to have radiological abnormalities reported than members of the general population. This could, and probably did, lead to earlier detection of the disease. However, we consider it to have had little effect on our estimations of survival as virtually all of the men were symptomatic before the first radiograph which showed irregular opacities. Other studies have also gauged onset by estimating the time when breathlessness was first noted by the patient (STACK, *et al.*, 1972; LIVINGSTONE, *et al.* 1964; TURNER WARWICK, *et al.*, 1980).

It may accordingly be considered that these men have IPF initially similar to that occurring in the general population, but which is in some way modified by the inhalation of coal dust. Comparable prospective studies from such populations might be informative.

This finding is not unique in Wales. The prevalence observed in a sample of West Virginia coal miners corresponds with previously published data for South Wales coal miners (Table 6) (COCKCROFT, 1985). There are no published data on the prevalence of IPF in post-mortem studies of the general population with which to compare our findings. The histological findings indicate that the overall prevalence at death in these populations of coal miners for moderate to severe forms of the disease is approximately 5%. Mild forms of the disease are discovered in another 10–12%. The *milder* forms are characterised by a relative lack of pigment and are not associated with emphysema. By contrast, the severe forms are associated with emphysema and appear to be extensions of pneumoconiotic lesions. They also appear to be more common in lungs showing silicotic granulomata.

TABLE 4. COMPARISON OF CLINICAL FEATURES

	Number	Mean age at onset years	Dyspnoea	Percent with clubbing	Percent with crackles	Airflow restriction	Reduced gas transfer
DILL <i>et al.</i> (1975)	11	47 (28-72)	Yes	54	82	80%	100%
LIVINGSTONE <i>et al.</i> (1964)	45	50.5 (20-78)	Yes	71	91	-	Yes
STACK <i>et al.</i> (1972)	96	-	Yes	-	100	-	95%
TUKIAINEN <i>et al.</i> (1983)	100	53 (16-77)	-	46	94	80%	Yes
TURNER-WARWICK <i>et al.</i> (1980)	220	54 ± 12 (18-84)	Yes	66	96	Yes	-
CARRINGTON <i>et al.</i> (1978)	130	40-50	-	-	60	Yes	Yes
SCADDING (1960)	26	53 (23-68)	Yes	85	100	0	Yes
CRYSTAL <i>et al.</i> (1976)	29	52	Yes	72	65	Yes	Yes
McCONNOCHIE <i>et al.</i>	45	55 ± 7 (41-70)	Yes	69	82	-	97%

TABLE 5. COMPARISON OF QUOTED SURVIVAL FIGURES

	Number	Mean length of illness in years from onset to death	Mean length of illness in years for survivors in study population	Mean length of illness in years for total study population	5 years survival	10 years survival	15 years survival
SCADDING (1960)	26	4.5*	5.0*	-	-	-	-
LIVINGSTONE <i>et al.</i> (1964)	45	2.1	5.79	4.0	31%	4%	-
STACK <i>et al.</i> (1972)	96	4.0	-	-	-	7.3%	-
DILL <i>et al.</i> (1975)	11	2.46	6.4	4.9	45%	18%	-
TURNER-WARWICK <i>et al.</i> (1980)	220	3.2	8.9	-	50%**	28%	-
TUKIAINEN <i>et al.</i> (1983)	100	-	-	-	50%	-	-
McCONNOCHIE <i>et al.</i>	45	11.4	7.9	7.5	84%	49%	18%

TABLE 6. INTERSTITIAL PULMONARY FIBROSIS AT AUTOPSY

National Coal-workers Autopsy Study (USA)	15.9%
South West Virginia Autopsy Population	17.8%
South Wales Post-mortem Study (COCKCROFT, 1985)	17.1%

Acknowledgements—We are grateful to Rochelle Althouse for statistical advice and to Dr. D. Jones of the Cardiff Pneumoconiosis Medical Panel.

REFERENCES

- CARRINGTON, C.B., GAENSLER, E.A., COUTU, R.E., FITZGERALD, M.X. and GUPTA, R.G. (1978) Natural history and treated course of usual and desquamative interstitial pneumonia. *N. Eng. J. Med* 298: 801–9.
- COCKCROFT, A.E. (1985) The importance of irregular opacities on the chest radiographs of coal-workers. *M.D. Thesis*, London University.
- COTES, J.E. (1979) Lung function assessment and application in medicine. Fourth edition, Oxford, Blackwell.
- CRYSTAL, R.G., FULMER, J.D., ROBERTS, W.C., MOSS, M.L., LINE, B.R. and REYNOLDS, H.Y. (1976) Idiopathic pulmonary fibrosis: clinical, histologic, radiographic, physiologic, scintigraphic, cytologic and biochemical aspects. *Ann Intern Med* 85, 769–88.
- DILL, J., GHOSE, T., LANDRIGAN, P., MACKEEN, A.D. and MACNEILL, A.R. (1975) Cryptogenic fibrosing alveolitis. *Chest* 67, 411–6.
- GOUGH, J. and WENTWORTH J.E. (1949) The use of tissue sections of entire organs in morbid anatomical studies *J.R. Micro.Soc.* 69, 231–33.
- INTERNATIONAL LABOUR OFFICE (1980) Classification of radiographs of the pneumoconioses. ILO Publications. Geneva.
- KLEINERMAN, J., GREEN, F., LAQUEUR, W., TAYLOR, G., HARLEY R., PRATT, P., WYATT, J. and NAEYE, R. (1979) Pathology standards for coalworkers' pneumoconiosis. *Archo, Patho and Labo Medo* 103, 375–429
- LIVINGSTON, J.L., LEWIS, J.G., REID, L. and JEFFERSON, K.E. (1964) Diffuse interstitial pulmonary fibrosis: a clinical, radiological and pathological study based on 45 patients. *Q J. Med* 33 71–103
- SCADDING, J.G. (1960) Chronic diffuse interstitial fibrosis of the lungs. *Br Med J* 1, 443–50.
- STACK, B.H.R., CHOO-KANG, Y.F.J. and HEARD, B.E. (1972) The prognosis of cryptogenic fibrosing alveolitis. *Thorax* 27, 535–42.
- TUKIAINEN, P., TASKINEN, E., HOLSTI, P., KORHOLA, O. and VALLE, M. (1983) Prognosis of cryptogenic fibrosing alveolitis. *Thorax* 38, 349–55.
- TURNER-WARWICK, M., BURROWS, B. and JOHNSON, A. (1980) Cryptogenic fibrosing alveolitis : Clinical features and their influence on survival. *Thorax* 35, 171–80.

DISCUSSION

T. R. P. GOFFE: What is the effect of cigarette-smoking on the production of PIF in view of the high rate of cigarette-smoking in your study group, – 44 out of 45 workers?

K. McCONNOCHE: I cannot comment on the effect of cigarette smoking on the production of PIF either in coal-workers or other members of the population. However, we know from the study of Turner-Warwick *et al.* (1980) that it has no effect on survival.

J. L. ABRAHAM: Are there any mineralogical data available yet on these cases?

K. McCONNOCHE: No.

E. MOORE: What data do you have concerning the coal-workers' primary occupation, their dust exposure, or the collieries at which they worked?

K. McCONNOCHE: Many of these workers have been exposed to stone and coal dust but the significance of this is unknown as we have not examined a control group. We have also collected data on collieries but have not yet found any associations.

J. E. BACHYNSKI: Radiographic patterns of Western Canadian coal-workers are irregular in the apices.

K. McCONNOCHIE: The Welsh and US workers start their radiographic changes in the bases.

A. G. HEPPELSTON: Do you consider the interstitial fibrosis is incidental to dust exposure or disease? Such fibrosis is seen in adults irrespective of occupation.

K. McCONNOCHIE: I think this pattern of interstitial fibrosis may be related to dust exposure though undoubtedly spontaneous cases will arise in coal-workers as in other adults. Our data suggest that the natural history in coal-workers is very different and we suspect that the prevalence is higher in this community than expected, though it would be difficult to prove.