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Fatal silicosis in a funeral arts' craftsman

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Parole chiave
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Summary
Background: *The current scientific debate on silica and cancer has often led to overlooking the persisting risk of lung fibrosis from crystalline silica.* **Case report:** *During 2000, when 54 years old, a funeral arts' craftsman began to suffer from persistent, hacking cough; radiography and high resolution computed tomography (HRCT) showed thin sub-pleural nodulations of the lungs, mainly in the upper fields. Two years later, increasing dyspnoea appeared and HRCT revealed vast consolidative opacities in both upper fields. Bronchial biopsies documented lung fibrosis and silicosis was diagnosed. In 2003, large amounts of crystalline silica were found in a stone used by the patient and in the dust deposited inside his workshop. In 2004 the patient abandoned his work. He died in 2008 from respiratory failure and infectious complications.* **Discussion:** *Histology is uncommonly available for the differential diagnosis of lung fibrosis so that, when imaging is not accompanied by appropriate recollection and assessment of occupational histories, new cases of silicosis may easily remain unrecognized. After some years from its onset, this fatal silicosis case showed a relatively rapid progression, that continued after exposure cessation. The disease derived from working conditions that the patient and his only colleague, both craftsmen operating in their own workshop, didn't recognize as hazardous prior to an external intervention.* **Conclusions:** *Severe and fatal silicosis is still present in Italy, even in unusual occupational contexts. At present the disease may be more difficult to identify than in the past; collaboration between clinicians, radiologists and occupational physicians is strongly needed. Ad hoc industrial hygiene surveys give a very relevant support to diagnosis and prevention of silicosis.*

Riassunto
«*Silicosi fatale in un artigiano delle arti funerarie*». **Introduzione:** *Il dibattito scientifico in corso su silice e cancro ha spesso distolto l'attenzione dalla persistenza di rischi di fibrosi polmonare derivanti dalla polvere di silice cristallina.* **Descrizione del caso:** *Nel corso del 2000, all'età di 54 anni, un artigiano delle arti funerarie iniziò a soffrire di tosse stizzosa persistente; radiografia e HRCT mostrarono una fine nodulazione sub-pleurica dei polmoni, principalmente nei campi superiori. Due anni più tardi comparve una dispnea ingravescente e la HRCT mostrò anche vaste opacità consolidative in entrambi i campi superiori. Biopsie bronchiali documentarono una fibrosi polmonare portando a classificare la malattia come una silicosi. Nel 2003, notevoli quantità di silice cristallina furono trovate in una*

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pietra usata dal paziente e nella polvere sedimentata all'interno del suo laboratorio. Nel 2004 il paziente abbandonò il suo lavoro. Egli morì nel 2008 per insufficienza respiratoria e complicanze infettive. **Discussione:** *L'istologia non è comune nella diagnosi della fibrosi polmonare cosicché, mancando un'integrazione tra storie lavorative appropriate e imaging, il riconoscimento di nuovi casi di silicosi rimarrà arduo. Dopo alcuni anni dall'insorgenza, questo caso di silicosi fatale mostrò una progressione relativamente rapida anche dopo la cessazione dell'esposizione. La malattia è derivata da condizioni di lavoro che il paziente e il suo unico collega, entrambi artigiani operanti in un laboratorio proprio, non avevano riconosciuto come pericolose prima che vi fosse un intervento esterno.* **Conclusioni:** *La silicosi grave e fatale è una criticità tuttora presente in Italia, anche in contesti lavorativi non classici. Oggi la malattia può essere più difficile da identificare rispetto al passato; un lavoro congiunto tra clinici, radiologi e medici del lavoro è necessario. Adeguati studi di igiene industriale forniscono un supporto di grande rilievo alla diagnosi e alla prevenzione della silicosi.*

Background

The current scientific debate on silica and cancer has often led to overlooking the persisting risks of lung fibrosis caused or, at least, triggered by crystalline silica dust. Certainly, in Western Europe quartz dust exposures are now less frequent and, on average, less intense than in the past, but this doesn't mean the risk of new silicosis cases, even in severe forms, has been eliminated.

Short but intense quartz exposures are always possible from occasionally uncontrolled work circumstances and disasters. Quartz dusts can develop in unusual work settings, where the hazard is more often unacknowledged and, therefore, neglected.

It has been hypothesized that autoimmune processes can be triggered and enhanced by silica dust, affecting respiratory tissues and others (8, 20, 27, 29). Under this assumption, silicosis occurrence and progression might partly result unpredictable. Silica might contribute to the occurrence and progression of other diseases. Silicosis and sarcoidosis can coexist (9, 10) and it has been argued that cases of lung sarcoidosis and pulmonary alveolar proteinosis (PAP) might be the outcome of peculiar reactions to crystalline silica (30). Increasing rates of progressive massive fibrosis are being reported in the United States, partly attributable to occupational silica exposure (23). This problem is probably not confined to the United States only and is probably understudied and underestimated in many countries.

Case presentation and investigations

An Italian man (born in 1946, never smoker, diagnosed with "type 2" diabetes mellitus in 1984, with no other main clinical conditions) always worked since adolescence as a funeral art's craftsman. Since 1973 his main task was engraving images and inscriptions upon marbles more frequently used as gravestones. As a minor activity, he machine sawed and sandblasted slabs of stone; sandblasting was carried out inside a cabin provided with partial enclosure and local exhaust. He progressively became a specialist in reproducing portraits from photographs on gravestones, sometimes deeply carving the marble, mostly "scratching" the images using a hand held, very thin electric milling cutter. Neither local exhaust nor respiratory protections were used during this operation; there was no technical possibility of wetting surfaces to abate dusts.

One of us (Ballatori) was the patient's general practitioner; being also an occupational health practitioner, in consideration of his job, even in absence of respiratory symptoms and radiographic signs of lung disease, in 1997 he performed a first screening spirometric test (Vitalograph Alpha® equipped with a Fleisch pneumotachograph) and detected a slight restrictive ventilatory failure (table 1).

During the summer of 2000, the patient began to suffer from persistent hacking cough and a plain chest radiography revealed thin sub-pleural nodulations of the lungs, mainly in the upper fields. A chest

Table 1 - Lung function tests from 1997 to 2002 (height of the subject: 176 cm; weight unrecorded) – the same Vitalograph Alpha® portable spirometer equipped with a Fleisch pneumotachograph was used for all measurements/tests

	FVC L/min	FEV1 L/min	FEV1/CV %	PEF L/min
1997 Nov 19	4.11	3.66	89	510
1998 Nov 18	4.02	3.58	89	516
1999 Nov 22	3.73	3.16	77	468
2000 Nov 09	3.27	2.84	82	354
2001 Dec 01	2.67	2.37	87	324
2002 Dec 02	1.79	1.23	59	84

high resolution computed tomography (HRCT) two months later confirmed this finding and revealed some thickening of the pleural fissures.

At successive spirometric tests from 1997 to 2002 the lung function parameters continued to decline (table 1).

In 2002 the patient began to suffer from increasing dyspnoea.

By the end of 2002 lung function tests documented worsening of the restrictive ventilatory pattern and a decrease in diffusing capacity of carbon monoxide (DLCO) (figure 1). A further chest HRCT revealed *"vast consolidative opacities in both the upper field*s" and lung cancer was suspected.

In January 2003 fibrobronchoscopy was performed and transbronchial lung biopsies were taken, showing: *"nodules constituted by concentric fibroblasts and many macrophages containing apparent granules at the polarized microscope"*. The pathologist's assessment was: *"histological pattern consistent with the diagnosis of silicosis"*. The attending pneumologist diagnosed a *"macronodular lung silicosis"* and reported the case as suspected occupational disease to the Occupational Health and Safety unit of the local health authority.

In February 2003 we visited the workshop where the patient had been working since 1983, with another coworker. They referred that the stones they used were represented by white Carrara marble (80%), by medium-textured black *"granitoids"* (8%), by a fine-textured black stone coming from Zimbabwe commercially defined as *"absolute black"* (2%) and by a mix of many other marbles and *"granitoids"* (10%). The grit used for sandblasting was plainly declared as quartz.

Carrara marble is almost pure calcium carbonate ($CaCO_3$) containing around 2% of other minerals, no more than a half of which consisting of crystalline silica (SiO_2) (5), so we did not further investigate on it. It is well known that granites and *"granitoids"* contain a high percentage (from 44 to 77% and more) of crystalline silica (12). We decided to study the *"absolute black"* stone and the abundant grey dust settled on every surface in the workshop and particularly around the sandblasting cabin. The sandblasting grit was loaded into the cabin and projected by compressed air through a hose handled by a single worker. On the front opening of the cabin some hanging plastic bands provided very limited protection against the release into the workshop of the dust not captured by the local exhaust system.

Samples of the *"absolute black"* stone and of settled dusts were taken and analysed for crystalline silica content by X-ray diffractometry (XRD). The *"absolute black"* stone quartz content was 13.4%. Settled dust was 96.3% quartz.

The patient's ventilatory function continued to decline and his dyspnoea to worsen. No serological signs of connective tissue diseases and other systemic diseases were ever found.

In March 2003 the patient underwent a second fibrobronchoscopy and bronchial biopsies. Histological examination showed micro-nodular fibrous foci diagnosed as silicotic lesions.

In March 2003, and in January 2004, further chest HRCTs documented two gross conglomerated fibrous masses in the upper fields (main diameter: 7 cm for the one in the left lung, 5 cm for the other in the right lung), diffuse sub-pleural micronodules in the upper and medium fields and in the lingula, thickening of the inter-lobular sects and enlargement of the hilar lymph-nodes.

In June 2004 the patient abandoned work (while the quartz grit was substituted with a quartz-free one).

In July 2004 plain chest radiography showed further bilateral progression of mixed small and large opacities and gross pleural thickening areas in both upper fields. The patient had dyspnoea after minimal efforts, such as a few steps on plain ground, and cough with very limited phlegm.

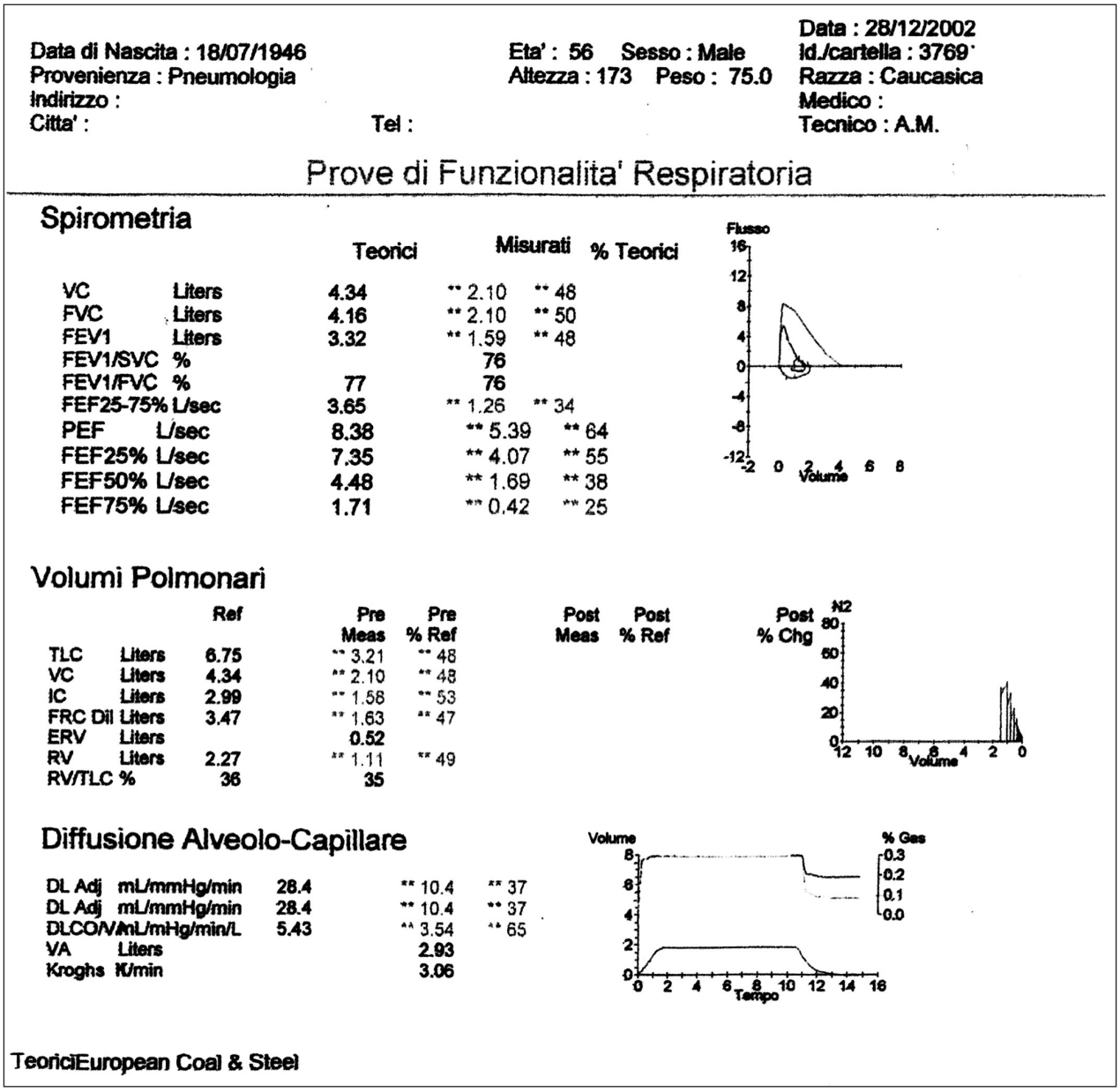

Figure 1 - Lung function test (28 December 2002)

Lung transplantation was ruled out. The patient died in 2008 from chronic lung failure complicated by bacterial infection and septic shock.

After the patient's death all of his personal documentation was lost, so that no radiograph and no lung function test performed since 2003 onwards is available for re-examination; we were not able to obtain slides or photographs of the patient's lung biopsies.

Discussion

Years ago, one of us (Calisti) and a colleague (Barbieri) described an Italian blue-collar worker who developed rapidly progressive fibrosis leading to lung transplantation. Histological examination of removed lungs showed diffuse fibroid aspects, interpreted by the pathologists as "sarcoidosis-like" or "silicosis-like" lesions. For five years this patient

worked at a Banbury mixer in a silicon rubber factory; he loaded into the mixer various raw materials, including quartz and cristobalite in powder (for use as fillers) and used to shovel the amount not incorporated into the rubber and fallen on the soil to reload it into the mixer. We discussed this case in a short communication presented in 2002 at a symposium about silica, silicosis, cancer and other diseases (2).

This further case of severe silicosis in Italy doesn't present glaringly peculiar features: it developed after years of work in dusty conditions, with slight symptoms at the start and, initially, without evidence of a tendency to worsening. The patient was certainly exposed to dusts from freshly cleaved quartz and it is well known that the inhalation of freshly cleaved quartz results in dramatic increases in all the pulmonary cytotoxic and inflammatory responses (24, 28).

On the other hand, this case deserves consideration for several reasons.

Severe silicosis is recognized as a critical, persisting issue in occupational health in developing countries, but we are no longer accustomed to observing fatal cases of silicosis in Western European countries. This case provides an alert to the possible reappearance of silicosis.

There certainly was long latency and initial slow progression, but since 2002 the patient's respiratory function deteriorated rapidly even if occupational dust exposures had ceased in 2004. It has been argued that when exposure to crystalline silica continues, the lung dust burden reaches an overload level where alternatively activated macrophages cannot accommodate further quartz particles, thus leading to progressive lung inflammation and fibrosis (11). Autoimmunity was supposed to at least partially explain sudden silicosis progression (8, 20, 27, 29). Biopersistence of crystalline silica in lung tissues has been evaluated by analysing the mineral content of broncho-alveolar lavage fluid (BALF); crystalline silica in BALF was shown to progressively decrease after exposure, without returning to the baseline even after ten or more years (19). Crystalline silica is thought to be a relevant agent in coal miners' lung disease, that can progress after exposure discontinuation (21). Several new silicosis cases appeared during a four-year follow-up of former denim sandblasters and radiographic progression was frequently observed among them (1). For high-risk subjects, medical surveillance has to be reasonably continued after exposure cessation.

Pleural involvement is unusually associated with pneumoconioses, except asbestosis, and this finding remains difficult to explain in the case presented here.

In clinical practice, microscopic examination of bioptic samples is not usually performed to investigate pulmonary fibroses. Based only on its radiological appearance, in absence of histology, this case could have been easily diagnosed as idiopathic pulmonary fibrosis (IPF) or sarcoidosis (4, 14, 16, 26), especially if the patient's occupational history had not been taken or evidence of silica exposure had been overlooked.

Both IPF (14) and sarcoidosis (18, 22, 31) have been associated with crystalline silica; silicosis shares immunological mechanisms with these pathological entities (6, 8). As a matter of fact, boundaries between silicosis, sarcoidosis and pulmonary alveolar proteinosis (PAP) appear now less defined than in the past (30) and one diagnosis does not exclude the others. A recent study on paediatric cases of sarcoidosis considered it as a systemic reaction triggered by inorganic dusts in predisposed individuals (17): a sort of pneumoconiosis.

We want to underline that silicosis will hardly be suspected if the attending physician is unaware that a patient was occupationally exposed to mineral dusts and appropriate examinations will not be carried out unless specific questions are addressed to radiologists, pneumologists and occupational physicians. In any case, when patients with occult or just suspected exposures are under investigation, a skilled knowledge of the imaging patterns of occupational lung diseases is especially critical for all these professionals' work (23).

Conclusions

This case was due to exposure outside obvious occupational settings, such as coal mining, stone quarrying, iron and steel industry, and pottery factories. Here silicosis is recognised as a medical and social issue, and practitioners are expected to suspect the

disease when a blue-collar worker complains of persistent hacking cough and increasing dyspnoea. Cohorts of blue-collar workers, furthermore, are often followed-up in the framework of surveillance programs dedicated to respiratory health.

Outside this contexts, a correct diagnosis of silicosis might be today more difficult than in the past, as the occupational history may not obviously suggest exposure to crystalline silica. The keyword in this case was "sandblasting". "Sandblasting" has been the common exposure of workers diagnosed with silicosis in unusual occupational settings, in recent years too all over the world: dental laboratory technicians in USA (3), glass etchers in Spain (15), denim sandblasters in Turkey (1, 7).

Further, progressive lung fibrosis might be currently difficult to recognize as silicosis because the disease is now expected to occur in mild forms: severe and fatal cases of silicosis could be labelled as IPF or sarcoidosis. Crystalline silica is a more common occupational hazards than currently believed: in a recent Australian cross-sectional survey 6.4% of respondent workers were exposed to respirable crystalline silica (3.3% at high level) (25). Ascertaining quartz-related exposures is mandatory for a correct interpretation of new cases of interstitial lung disease and consequently for the correct treatment of these patients (13). It is as well essential for prevention, as these cases may be seen as "sentinel events". Collaboration between clinicians and occupational physicians is strongly needed.

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References

1. Akgun M, Araz O, Ucar EY, et al: Silicosis appears inevitable among former denim sandblasters: a 4-year follow-up study. Chest 2015; 148 (3): 647-654
2. Barbieri PG, Calisti R: About one case of acute silicosis: a current risk in the rubber industry. In Sun Y, Voss B (eds): Proceedings of the 3° International Symposium on Silica, Silicosis, Cancer and Other Diseases. Santa Margherita Ligure, 21-25 Oct. 2002. Med Lav 2002; 93 (suppl): S67-S68
3. Centers for Diseases Control and Prevention (CDC): Silicosis in dental laboratory technicians – five states, 1994-2000. MMWR Morb Mortal Wkly Rep 2004; 12 (53): 195-197
4. Chapman JT: Interstitial lung disease, 2010. Available online at the address www.clevelandclinicmeded.com/medicalpubs (last accessed 23 August 2016)
5. Criscuolo A, Lisi S: Geology and structures of marble formation. In Blasi P, Criscuolo A, Lisi S (eds): Proceedings of the Workshop The Carrara marble: geological, economic and mineralogical aspects. Carrara, 12-16 Sept 1998. Carrara: Eurominerals and the Society of Mining Professors, 1998: 9-13 (Article in Italian)
6. Davis GS, Holmes CXE, Pfeiffer LM, Hemenway DR: Lymphocites, lymphokines, and silicosis. J Environ Pathol Toxicol Oncol 2001; 20 (suppl 1): 53-65
7. Doganay S, Gocmen H, Yikilmaz A, Coskun A: Silicosis due to denim sandblasting in young people: MDCT findings. Eurasian J Med 2010; 42 (1): 21-23
8. Huaux F: New developments in the understanding of immunology in silicosis. Curr Opinion Allergy Clin Immunol 2007; 7 (2): 168-173
9. Hubener E, Kuhne W, Scharkoff T: (Coincidence of silicosis and sarcoidosis. 1 Observations in a selected patient sample). Z Erkr Atmungsorgane 1986; 165 (2): 130-133 (Article in German)
10. Hubener E, Kuhne W, Scharkoff T: (Coincidence of silicosis and sarcoidosis. 2 Relations between silicosis and sarcoidosis as well as forensic consequences). Z Erkr Atmungsorgane 1986; 166 (2): 186-193 (Article in German)
11. Kawasaki H: A mechanistic review of silica-induced inhalation toxicity Inhal Toxicol 2015; 27 (8): 363-377
12. Le Maitre RW, Bateman P, Dudek A, et al: A classification of igneous rocks and glossary of terms: recommendations of the International Union of Geological Sciences Subcommission on the Systematics of Igneous Rocks. Oxford (UK) Blackwell Scientific Publications, 1989
13. Leung CC, Yu IT, Chen W: Silicosis. Lancet 2012; 26: 379 (9830): 2008-2018
14. Meltzer EB, Noble PW: Idiopathic pulmonary fibrosis. Orphanet J Rare Dis 2008; 26: 3:8
15. Mendez-Vargas MM, Soto-de la Fuente AE, Soto-Vera EA, Leo-Méndez R: Acute silicosis. An infrequent pneumoconiosis. Rev Med Inst Mex Seguro Soc 2015; 53 (4): 524-527 (article in Spanish)
16. Meyer KC: Diagnosis and management of interstitial lung disease. Transl Respir Med 2014; 13: 2:4
17. Nathan N, Cavalin C, Macchi O, et al: Inorganic exposome in pediatric sarcoidosis: The PEDIASARC study. Europ Resp J 2015; 46 (suppl 59): PA849

18. Newman KL, Newman LS: Occupational causes of sarcoidosis. Curr Opin Allergy Clin Immunol 2012; 12 (2): 145-150
19. Pairon JC, Billon-Galland MA, Iwatsubo Y, et al: Biopersistence of nonfibrous mineral particles in the respiratory tracts of subjects following occupational exposure. Environ Health Perspect 1994; 102 (suppl 5): 269-275
20. Parks CG, Conrad K, Cooper GS: Occupational exposure to crystalline silica and autoimmune disease. Environ Health Perspect 1999; 107 (suppl 5): 793-802
21. Petsonk EL, Rose C, Cohen R: Coal mine dust lung disease. New lessons from old exposures. Am J Respir Crit Care Med 2013; 187 (11): 1178-1185
22. Rafnsson V, Ingimarsson O, Hjalmarsson I, Gunnarsdottir H: Association between exposure to crystalline silica and risk of sarcoidosis. Occup Environ Health 1988; 55 (10): 657-660
23. Seaman DM, Meyer CA, Kanne JP: Occupational and environmental lung disease. Clin Chest Med 2015; 36 (2): 249-268
24. Shoemaker DA, Pretty JR, Rameau DM, et al: Particle activity and in vivo pulmonary response to freshly milled and aged alpha-quartz. Scand J Work Environ Health 1995; 21 (suppl 2): 15-18
25. Si S, Carey RN, Reid A, et al: The Australian Work Exposures Study: prevalence of occupational exposure to respirable crystalline silica. Ann Occup Hyg 2016; 60 (5); 631-637
26. Spagnolo P, Sverzellati N, Wells AU, Hansell DM: Imaging aspects of the diagnosis of sarcoidosis. Eur Radiol 2014; 24 (4): 807-816
27. Steenland K, Goldsmith DF: Silica exposure and autoimmune diseases. Am J Ind Med 1995; 28 (5): 603-608
28. Vallyathan V, Castranova V, Pack D, et al: Freshly fractured quartz inhalation leads to enhanced lung injury and inflammation. Potential role of free radicals. Am J Respir Crit Care Med 1995; 152 (3): 1003-1009
29. Vigliani EC, Pernis B: An immunological approach to silicosis. J Occup Med 1959; 1 (6): 319-328
30. Vincent M, Chemarin C, Cavalin C, et al: From the definition of silicosis at the 1930 Johannesburg conference to the blurred boundaries between pneumoconioses, sarcoidosis, and pulmonary alveolar proteinosis (PAP). Am J Ind Med 2015; 58 (suppl 1): 31-38
31. Yeager H, Gopalan S, Mathew P, et al: Sarcoidosis: can a murine model help define a role for silica? Med Hypotheses 2012; 78 (1): 36-38

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The 7th ICOH International Conference on Work Environment and Cardiovascular Diseases. May 3-5, 2017, Varese, Italy

The ICOH Scientific Committee on Cardiology (SCC) in Occupational Health is a committee of researchers, cardiologists, occupational health physicians and public health workers throughout the world concerned about the impact of work and work-related factors on cardiovascular health. The SCC works to link economic globalization to the changing nature of work and its impact on the risk of cardiovascular diseases, and focuses on research, worksite interventions, and public health.

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