



DEPARTMENT OF VETERANS AFFAIRS
Veterans Health Administration
Washington DC 20420

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UNDER SECRETARY FOR HEALTH'S INFORMATION LETTER

SARCOIDOSIS

1. Background

a. Sarcoidosis among United States (U.S.) Navy personnel has been of interest since the mid 1940s, with publications addressing frequency, diagnostic criteria, and potential linkage with military service. Sarcoidosis is considered a multi-system disorder of unknown etiology characterized by noncaseating granulomas with pulmonary involvement in 90 percent of cases. Point prevalence in the U.S. civilian population is estimated at 10-20 per 100,000, with aggregation along geographic, familial, and racial lines. African-Americans have a 2.4 percent lifetime risk, individuals of Swedish descent, and other Caucasian descent, have a 0.85 percent lifetime risk. The condition probably represents an immunologically mediated pathological response to an environmental agent or agents in vulnerable individuals, with as yet poorly defined immunogenetics. The frequency has been declining slowly among U.S. Navy veterans, from greater than 5 cases per 100,000 before 1975 to 3 cases per 100,000 in 2001, with a steeper decline among African-Americans than among individuals of Caucasian descent.

b. In 1999, following a publication on sarcoidosis in Navy personnel, concern was expressed by some veterans and their advocates that the diagnosis of sarcoidosis in a Navy deck grinder was a misdiagnosis of pneumoconiosis. An Information Letter (IL 10-99-004) was published and a review of selected Department of Veterans Affairs (VA) cases did not confirm any widespread misdiagnosis. Still, some cases diagnosed as well-established sarcoidosis might actually reflect silicosis or mixed-dust pneumoconiosis. No data are available to determine whether sarcoidosis initially diagnosed long after separation from service actually reflects some other disease that is misdiagnosed as, or evolves into, sarcoidosis.

2. Epidemiology and Pathogenesis. Meanwhile, cohort and case-control studies of sarcoidosis suggest specific potential causes for at least some portion of cases diagnosed as sarcoidosis. Specific job categories (Navy Enlisted Classifications) are at higher risk in several cohort studies. Deck-grinding in the U.S. Navy, with silica exposure represents one such risk, although that specific activity showed no elevated risk in a follow-up study. Several additional job categories appeared at risk in a follow-up study including ship's servicemen, mess management specialists, and aviation structural mechanics: each with relative risks of less than 2 to 1. Recent case-control studies suggested that exposure to silica and indoor environments presented a minimally, though statistically significantly, increased risk for sarcoidosis. With one notable exception, odds ratios were relatively low, generally below 2 to 1. A histopathology and microanalytical study of a small, non-random sample of Navy veterans documented exposure to inorganic particulates matching the components found in non-skid paints (including crystalline

silica, talc, aluminum silicates, titanium dioxide, aluminum, and other metals) in the lungs of Navy sarcoidosis patients. The combination of exposures, including silica, titanium and other metals, suggests the presence of mixed-dust pneumoconiosis, a generally non-progressive disorder, rather than silicosis. Although silica exposure, rather than silicosis, is thought to present a risk factor for lung cancer, such low-grade exposures are not associated with the development of malignancy.

3. Pathology. Pulmonary lesions are thought to arise from an initial alveolitis involving interstitial infiltration of inflammatory cells (monocytes, macrophages and lymphocytes) which then evolves into the classical noncaseating granulomas. Although the nodules of mature silicosis, with dense, whorled fibrosis, differ substantially from those of sarcoidosis, which in rare cases resemble the less well-formed granulomas of hypersensitivity pneumonitis, some researchers consider that early changes may be similar and represent simply a non-specific alveolitis, indistinguishable from many other forms of interstitial pneumonitis. In general, the clinical course of silicosis and mixed-dust pneumoconiosis, as studied over many years, generally reflects very slow progression or simple persistence of radiographic abnormalities without progressive pulmonary disability. Where such progressive disease ensues, characteristic radiographic changes, including small to medium rounded opacities, hilar “egg shell calcification,” and progressive massive fibrosis occur. Progressive sarcoidosis related to bioaerosols, moisture, and indoor environments may represent ongoing exposure to an active agent, such as bioaerosols, and should lead to occupational and environment history taking focused on the current home and work environment.

4. Treatment. The clinical treatment of sarcoidosis after removal from exposure to possible inciting agents, with anti-inflammatory agents including steroids and cytotoxic drugs and oxygen, will not differ because of underlying causes in the distant past. No evidence supports the use of lavage, which has been proposed for early dust-induced disease.

5. Diagnosis and Compensation and Pension Examinations. When Navy veterans present for compensation and pension examinations, clinicians should obtain a detailed medical, occupational and environmental history focusing on silica, other inorganic dusts, and bioaerosols exposure. Clinicians should determine the initial onset of symptoms or diagnosis consistent with sarcoidosis.

a. In addition, they should identify the pertinent Navy Exposure Classifications. These include Ship’s servicemen, Mess management specialists, and Aviation structural mechanics. Clinicians should also obtain a description of the specific tasks, including deck-grinding and sand-blasting of paint, or proximity to such activities, such as serving in the hangar bay. Chest x-rays should be examined for traditional markers of sarcoidosis, silicosis, or mixed dust pneumoconiosis. If an exposure history suggests the presence of silica or other dust exposure, chest x-rays should also undergo scrutiny for the presence of dust-induced disease, using the classification system for B-readings of the American College of Radiology and the International Labor Organization. The presence of small rounded opacities in the upper zones of the lung may raise suspicion of silicosis. True egg-shell calcification in the hilar nodes, similarly, is considered suggestive.

b. When sarcoidosis arises during service, it represents a service-connected disease. The degree of disability follows standard assessment strategies as defined in the Automated Medical Information Exchange (AMIE) worksheet 1515, pulmonary interstitial and restrictive disease, listed under tuberculosis. No presumption of service-connectedness exists for sarcoidosis, a common disease, when it arises after separation from service.

6. References. See references and other pertinent material at the following Environmental Agents Service (131) websites: <http://www.va.gov/environagents> and <http://vaww.va.gov/vironagents> .

7. Inquiries. Questions concerning this information letter should be addressed to Michael Hodgson, MD, MPH, Occupational Health Program (136); 810 Vermont Avenue, NW; Washington, DC 20420.

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