



**DEPARTMENT OF VETERANS AFFAIRS  
Veterans Health Administration  
Washington DC 20420**

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In Reply Refer To: 13

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**UNDER SECRETARY FOR HEALTH'S INFORMATION LETTER**

**SARCOIDOSIS**

**1. Background**

a. Sarcoidosis is an idiopathic, multi-system disorder, which is characterized by noncaseating granulomas. Although the specific causes of sarcoidosis have not been confirmed, this disease is thought to result from a complex interaction of genetic factors and the environment. Family clusters of sarcoidosis are common. Possible precipitating causes among susceptible individuals include exposure to:

- (1) Environmental chemicals,
- (2) Viruses,
- (3) Bacteria,
- (4) Molds, and
- (5) Certain industrial dusts.

b. Although sarcoidosis can occur in almost any part of the body, it most often affects the lungs. Pulmonary involvement occurs in 90 percent of cases. Sarcoidosis frequently is asymptomatic, or mildly symptomatic, and may improve spontaneously without medical treatment. In many patients, sarcoidosis is diagnosed because of abnormal findings on a routine chest X-ray; however, the clinical course of sarcoidosis may also worsen over time.

c. Sarcoidosis affects people of all ages and races worldwide. It is more common in adults between 20 and 40 years of age and slightly more common in women than in men. Point-prevalence of sarcoidosis in the United States (U.S.) civilian population is estimated at 10 to 20 per 100,000. Of particular note, sarcoidosis is at least three times more common in African-Americans than in other racial and/or ethnic populations, and the disease course tends to be more acute and severe in African-Americans. The lifetime risk of developing sarcoidosis is about 1 percent among Caucasians and 2.4 percent among African Americans.

## 2. Diagnosis and Clinical Management

a. Sarcoidosis often causes no noticeable health problems. In symptomatic patients, the level of disease is directly related to the effects of granuloma inflammation on bodily functions. When granulomatous involvement is sufficient to cause disease, sarcoidosis typically presents with fatigue, feverishness, weight loss, shortness of breath on exertion, and a dry cough. In acute symptomatic disease, patients may also present with polyarthralgias, erythema nodosum, and bilateral hilar lymphadenopathy.

b. Sarcoidosis can affect any organ system, but most commonly, the lungs, heart, liver, eyes, skin, lymphatic system, nervous system, and kidneys. Both thoracic and peripheral lymphadenopathy are frequently observed. Anterior uveitis is also common and typically presents with photophobia, blurred vision, and tearing.

c. Disease caused by sarcoidosis often remits; however, the clinical course of sarcoidosis may be slowly progressive. Sarcoidosis can less frequently have an acute, rapidly advancing clinical course. Sarcoidosis can also cause death from granulomatous inflammation. The two most common causes of death are progressive pulmonary fibrosis and cardiac involvement.

d. Sarcoidosis can be a difficult disease to diagnose because its clinical presentation is so varied, the etiology is unknown, and there is no diagnostic test commercially available. Sarcoidosis, therefore, has to be diagnosed by confirming the presence of noncaseating granulomas and by excluding other known causes of granulomatous inflammation. Several types of medical tests usually are needed to diagnose sarcoidosis. A chest X-ray showing hilar adenopathy is suggestive. Confirmation of sarcoidosis requires biopsy, most often a transbronchial lung biopsy or skin biopsy.

e. Sarcoidosis can be mistakenly diagnosed for other known causes of granulomatous inflammation and other chronic lung diseases. The differential diagnosis includes hypersensitivity pneumonitis, mycobacterial infection, fungal infection, beryllium exposure, and exposure to metal dusts or organic antigens. Therefore, additional tests need to be considered when noncaseating granulomas are found by a biopsy including:

- (1) Antineutrophil cytoplasmic antibodies for Wegener's granulomatosis,
- (2) Anti-mitochondrial antibodies for primary biliary cirrhosis,
- (3) Serologic and skin tests for fungal infections,
- (4) Sputum culture for mycobacteria, and
- (5) Beryllium lymphocyte proliferation test for beryllium exposure.

f. Chest radiographs need to be examined for traditional markers of silicosis and mixed-dust pneumoconiosis, as well as for sarcoidosis. If an exposure history suggests the presence of silica or other dusts, chest X-rays need to be scrutinized for the presence of dust-induced disease using

the B-reading classification systems of the American College of Radiology (ACR) and the International Labor Organization (ILO). 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 is also considered suggestive of silicosis.

g. Drug therapy is available for symptomatic sarcoidosis, when needed. Sarcoidosis is usually treated initially with corticosteroids. Cytotoxic drugs (like methotrexate) and hydroxychloroquine may also be effective. Other non-specific types of therapies may be helpful, depending on the type of functional impairment caused by the disease. No currently available evidence supports the use of lavage for sarcoidosis, as has been proposed for early dust-induced disease.

### **3. Studies of Sarcoidosis Among Military Populations**

a. The U.S. Navy has been studying sarcoidosis since the 1940s. Recent research by the Navy and the Centers for Disease Control and Prevention (CDC) identified a higher-than expected number of sarcoidosis cases in military personnel who served aboard aircraft carriers. As part of the on-going effort to learn more about sarcoidosis, the Naval Health Research Center, San Diego, CA, and the Armed Forces Institute of Pathology (AFIP), Washington, DC, studied reported cases of sarcoidosis among Navy enlisted personnel from 1975 to 2001. They evaluated possible relationships between shipboard occupational duties and hospitalization for sarcoidosis. The final report of the Navy Lung Disease Assessment Program was published in February 2004. Study findings included:

- (1) Declining numbers of diagnosed sarcoidosis cases in the Navy since the mid-1970s,
- (2) Increased rates among African-Americans,
- (3) Increased rates in certain groups of enlisted personnel who served on aircraft carriers, including laundry and dry cleaning operators, barbers, mess management specialists, and aviation structural mechanics,
- (4) Increased incidence in specific Navy enlisted job classifications, and
- (5) Possible association between disease and exposure to deck-grinding abrasives.

b. In contrast to this last finding, the risk of sarcoidosis was not elevated in some job categories that would typically be associated with deck work or deck departments, i.e., those groups likely to perform deck grinding activities, such as Boatswain's Mates (BM), Aviation Boatswain's Mates (ABM), and general duty seamen. Excess rates among some occupational groups may have been due to spurious, chance findings from multiple statistical testing. Additionally, the finding of an increased risk of sarcoidosis in aircraft carriers may have been confounded by the fact that larger ships have better-equipped medical facilities, which typically perform more X-rays, increasing the likelihood of identifying sarcoidosis among crew members.

c. In addition to the possibility of chance findings, a histopathology and micro-analytical study of pathology specimens obtained from a small, non-random sample of Navy veterans suggested a potential increase in the presence of inorganic particles matching the components found in non-skid paints (including crystalline silica, talc, aluminum silicates, titanium dioxide, aluminum, and other metals) in the lungs of a few Navy patients diagnosed with sarcoidosis. However, this analysis did not reach statistical significance. Lastly, data are not available to determine whether veterans who were diagnosed with sarcoidosis after separation from active duty developed the initial stages of the disease during military service.

d. As noted, sarcoidosis is not unique to the military. Civilians in all walks of life develop sarcoidosis. Non-military research studies have tried to identify if certain exposures are associated with the development of disease. Studied exposures have included:

(1) Mold (often related to moisture, that which occurs in shower stalls and in damp basements);

(2) Agricultural work (including livestock handling);

(3) Insecticides;

(4) Building hardware (i.e., wood products and mobile home manufacturing); and

(5) Industrial dusts.

e. None of these exposures has been conclusively demonstrated to cause sarcoidosis. Unexpectedly, teachers have been found to be at increased risk of developing sarcoidosis.

f. In summary, studies of the etiology of sarcoidosis in civilian populations and among Navy personnel have been inconclusive and have not answered important questions as to whether sarcoidosis is caused by specific occupational and environmental exposures during military service.

#### **4. Outreach and Clinical Care**

a. The Department of the Navy, in coordination with the Department of Veterans Affairs (VA), is conducting an outreach program to ensure that service members and veterans with sarcoidosis are informed of possible long-term health problems. This Information Letter is part of this effort and is intended to help health care providers and disability specialists in the evaluation of veterans who may have developed sarcoidosis.

b. For military personnel who are still on active duty and have health concerns about chronic lung disease, they need to be directed to contact their nearest military health care provider.

c. For veterans who have left active military service and have concerns about chronic lung disease, they may be eligible to enroll for VA health care. Veterans with sarcoidosis do not currently have special eligibility for VA health care benefits. However, if VA determines that

their sarcoidosis is service-related, they are eligible for cost-free VA health care for service-related disabilities. Veterans need to be encouraged to visit VA's health eligibility website for further information, <http://www.va.gov/healtheligibility/> or call the VA Health Revenue Center toll free number, 1-877-222-8387.

d. When veterans present for evaluation of chronic lung disease, clinicians need to obtain a detailed medical and occupational and/or environmental history focusing on exposures to silica, other inorganic dusts, and bioaerosols. Clinicians need to determine when symptoms initially began and whether the diagnosis is consistent with sarcoidosis. In addition, they need to identify the pertinent military job descriptions and obtain a description of the specific tasks, including deck-grinding and sand-blasting of paint, or proximity to such activities. Non-military occupational exposures to silica, other inorganic dusts, and bioaerosols also need to be identified and recorded. To aid in the collection of this information, VA Form 10-0445, Occupational and Environmental Exposure History, can be found on the Intranet at <http://vaww.va.gov/vaforms/medical/pdf/vha-10-0445-fill.pdf>, or the Internet at <http://www.va.gov/vaforms/medical/pdf/vha-10-0445-fill.pdf>. This information may be useful for determining the cause of chronic lung problems among veterans and may be useful for exploring potential causal relationships between various exposures and sarcoidosis.

e. For veterans who suffer from chronic health problems and who think their illness is related to military service, they can contact VA toll free at 1-800- 827-1000 to speak to a VA representative about filing a disability claim. They may also file a claim on-line by going to <http://vabenefits.vba.va.gov/vonapp/main.asp>.

f. If physical impairment is found to be caused by sarcoidosis, entitlement to service-related disability compensation may be established if sarcoidosis is diagnosed during military service or within 1 year following discharge from active duty. When sarcoidosis is diagnosed more than 1 year following discharge from the military, entitlement to service-related disability compensation may also be established on an individual basis if medical evidence establishes a link between current disability and military service.

g. The assessment of the degree of disability due to sarcoidosis follows standard assessment strategies as defined in VA's Automated Medical Information Exchange (AMIE) worksheet 1515, Pulmonary Interstitial and Restrictive Disease, listed under Tuberculosis. Refer to: <http://vbaw.vba.va.gov/bl/21/rating/medical/exams/disexm49.htm>.

## 5. References

a. Environmental Agents Service (131) web site: <http://www.va.gov/environagents>.

b. Abraham, JL and Panitz, E., "Is "Sarcoidosis" in the U.S. Navy Occupational Lung Disease from the Grinding of Non-Skid Paint Aboard Aircraft Carriers? Analysis of Inorganic Particulates in Lungs and a Reference Paint Sample," American Journal of Respiratory Critical Care Medicine, 163:A214, 2001.

c. Gorham et. al., "Trends and Occupational Associations in Incidence of Hospitalized Pulmonary Sarcoidosis and Other Lung Diseases in Navy Personnel," Chest. 126:1-8, 2004.

d. Jajosky, P. "Sarcoidosis Diagnoses Among U.S. Military Personnel: Trends and Ship Assignment Associations," American Journal of Preventive Medicine. 14 (3): 176-83; Apr. 1998.

e. Newman, LS, Rose, CS, Maier, LA. "Sarcoidosis," New England Journal of Medicine. 336:1224; 1997.

f. Rybicki BA, Major M, Popovich J Jr., et al. "Racial Differences in Sarcoidosis Incidence: A 5 Year Study in a Health Maintenance Organization," American Journal of Epidemiology. 145: 234; 1997.

g. Seaton, A, Seaton, D, Leitch, AG. (Editors) Crofton and Douglas's Respiratory Diseases. Fifth Edition. Blackwell Science Ltd. Oxford. 2000.

**6. Inquiries.** Questions concerning this Information Letter need to be addressed to Dr. Kenneth Hyams, Office of Public Health and Environmental Hazards (13); 810 Vermont Avenue, NW; Washington DC 20420. The telephone number is 202-273-8579.

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