

Does sarcoidosis run in families?



Yes, Sarcoidosis does run in families. The ACCESS study found that the risk for sarcoidosis was increased nearly 5-fold in parents and siblings of patients with the disease. Familial aggregation was much more prominent in whites than in African-Americans, but an important risk factor in both races

Prof. Robert P. Baughman, of the University of Cincinnati, said **"We had a substantial number of white patients with severe disease. This finding challenges the widely held stereotype that the patients most often affected are African-Americans and young adults."***

The analysis found that sarcoidosis does not primarily affect young adults. **Half of the patients were older than 40 years at the time of diagnosis and some were in their 60s and 70s.**

The disease had already affected one organ in 50% of these newly-diagnosed patients, two organs in 30%, while 20% had three or more organs involved. The lungs were most commonly affected, in 95% of all patients. The skin was affected in 16%, the lymph nodes, in 15%, and eyes and liver, in 12% each.



Can my spouse get sarcoidosis?

The ACCESS study noted that there were five husband-and-wife couples who both had sarcoidosis.** Yet sarcoidosis is such a rare disease that, based on statistics, there should have been none. Perhaps this is weak evidence of communicability, or maybe it indicates that shared experiences increase the risk factors. Nevertheless, It would seem wise to monitor the health of spouses.



From:
Autoimmunity Research Foundation Inc,
www.AutoimmunityResearch.org
3423 Hill Canyon Ave,
Thousand Oaks, CA 91360 USA



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SARCOIDOSIS

Lessons Learned from the NIH ACCESS Study



* Begany T: "New data alter view of sarcoidosis"
PulmonaryReviews.com, Vol.7, No.4

** "Familial aggregation of sarcoidosis" Am J Respir Crit
Care Med. 2001 Dec 1;164(11):2085-91

ACCESS found that the Majority of Sarcoidosis Patients Do Not Get Better During the First Two Years, and Noted that Very Few Get Better After That

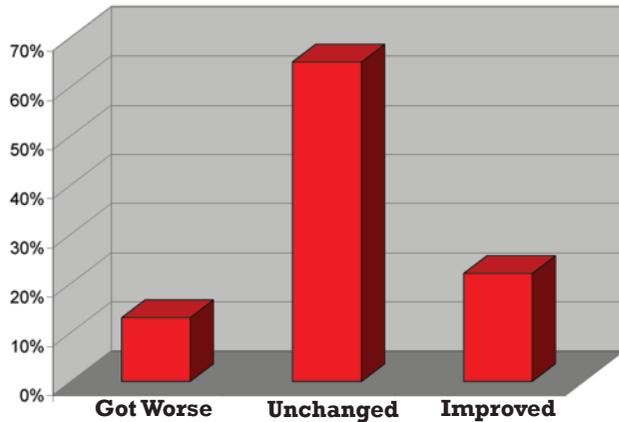


Figure 1: At the two-year follow-up, data from all patients was collated together, including the patients who had been taking prednisone. Only 22% showed a measurable improvement, and improvement "was not statistically related to whether the patients did or did not receive therapy".*

What was the ACCESS Study?

The ACCESS study represented "a serious effort to gain new understanding of both the cause and course of sarcoidosis".**

It was conducted by the National Heart, Lung and Blood Institute (NHLBI) of the National Institutes of Health (NIH) during the 6 year period 1995-2001.

Ten major U.S. university clinical centers took part in the study, at a cost of tens of millions of dollars. They followed 215 newly diagnosed patients for two years, the period within which it is thought that Sarcoidosis might go into remission.

Nevertheless, unless you have access to a good medical library, the study results are hard to locate. The Autoimmunity Research Foundation has created this brochure to gather together the information most vitally important to Sarcoidosis

patients, their families, and their physicians.

The Results of the ACCESS Study

The study found that the measures of Sarcoidosis severity: "FVC, FEV1, Scadding stage, and the dyspnea scale, remained unchanged over the two-year period in the majority of the patients".*

Further, it was noted "most patients with persistent disease at two years were unlikely to have resolution of Sarcoidosis" and "end-stage pulmonary Sarcoidosis usually develops over one or two decades".*

"In addition, the use of corticosteroids may promote relapse of Sarcoidosis when the medication is discontinued".*

"50 of the 215 (23%) of patients developed one or more new organ involvement over the two-year follow-up period".*

Surprises

- There were no documented cases of 'spontaneous resolution'.
- The use of corticosteroids by some of the study cohort actually made their sarcoidosis worsen.

In fact, the investigators concluded "the interaction of corticosteroid therapy with two-year outcome is complex .. More patients with improved FVC or dyspnea at follow-up were taking corticosteroids than not, and more patients with a worse FVC or dyspnea at follow-up were taking corticosteroids than not".*

"Patients taking corticosteroids tended to be improved or worsened with respect to FVC and dyspnea more often than those not taking corticosteroids".*

Issues still to be addressed

"ACCESS did not obtain data concerning the dose of corticosteroids that was used by patients, which may also affect the rate of relapse".*

"Another limitation of the ACCESS analysis of chest radiographs is that the initial and follow-up chest radiographs were not directly compared".*

Understanding Terms Doctors May Use

(Taken from the Johns and Michele paper**)

Sarcoidosis outcome can be documented as 'spontaneous resolution', 'improvement', 'worsening', 'unchanged', or 'indeterminate'.

"**Spontaneous resolution** is complete clearing of disease for at least 1 year without treatment."

"The **improvement** category includes patients either on or off therapy with decreased intra- or extrathoracic disease without worsening of the other, whereas worsening includes patients with increased intra- or extrathoracic disease without improvement in the other."

"An **unchanged** status means that the patient has had no improvement or worsening while off treatment for 1 year, while **indeterminate** status includes patients who do not fit into one of the other 4 categories above. "

"Other terms are often used in the literature such as '**spontaneous remission**,' which is a measurable decrease in degree or extent of disease that occurs without specific treatment. Remissions may be either partial or complete. A '**relapse**' represents worsening that occurs after a significant period of improvement either on or off treatment, usually following tapering or discontinuation of treatment".

The Autoimmunity Research Foundation

(www.AutoimmunityResearch.org)

ARF is a charity founded by recovered patients, patient advocates, and medical professionals.

We are dedicated to "solving the enigma of 'autoimmunity', one disease at a time." Our volunteer research staff has published numerous papers, and is actively engaged in internet-based studies and outreach. If you are able to send a donation to help us in our work, you can use our website to donate funds from a credit card, or you can send a check to the address on the back of this brochure.

This brochure was prepared by our Scientific Committee: Trevor G Marshall, PhD; Meg Mangin, RN; and Belinda Fenter, BS.

* The ACCESS Group: Two year prognosis of sarcoidosis: the ACCESS experience. Sarcoidosis Vasc Diffuse Lung Dis. 2003 Oct;20(3):204-11

**Johns CJ, Michele TM: The clinical management of sarcoidosis. A 50-year experience at the Johns Hopkins Hospital. Medicine (Baltimore). 1999 Mar;78(2):65-111.

A hyperlinked analysis of the report: "Two year prognosis of sarcoidosis: the ACCESS experience" is available from <http://AutoimmunityResearch.org/access-2yr.htm>