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Clinical phenotypes of extrapulmonary sarcoidosis: an analysis of a French, multi-ethnic, multicentre cohort

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There are five distinct phenotypes of sarcoidosis, with a non-random distribution of organ involvement. These five phenotypes differ according to sex, geographical origin and socioprofessional category. <https://bit.ly/3iCurZK>

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ABSTRACT Sarcoidosis is a rare disease of unknown cause with wide heterogeneity in clinical features and outcomes. We aimed to explore sarcoidosis phenotypes and their clinical relevance with particular attention to extrapulmonary subgroups.

The Epidemiology of Sarcoidosis (EpiSarc) study is a French retrospective multicentre study. Sarcoidosis patients were identified through national hospitalisation records using appropriate codes from 11 hospital centres between 2013 and 2016 according to a standardised protocol. Medical charts were reviewed. The phenotypes of sarcoidosis were defined using a hierarchical cluster analysis.

A total of 1237 patients were included (562 men and 675 women). The mean age at sarcoidosis diagnosis was 43.5±13 years. Hierarchical cluster analysis identified five distinct phenotypes according to organ involvement and disease type and symptoms: 1) erythema nodosum, joint involvement and hilar lymph nodes (n=180); 2) eye, neurological, digestive and kidney involvement (n=137); 3) pulmonary involvement with fibrosis and heart involvement (n=630); 4) lupus pernio and a high percentage of severe involvement (n=41); and 5) hepatosplenic, peripheral lymph node and bone involvement (n=249). Phenotype 1 was associated with being European/Caucasian and female and with non-manual work, phenotype 2 with being European/Caucasian, and phenotypes 3 and 5 with being non-European/Caucasian. The labour worker proportion was significantly lower in phenotype 5 than in the other phenotypes.

This multicentre study confirms the existence of distinct phenotypes of sarcoidosis, with a non-random distribution of organ involvement. These phenotypes differ according to sex, geographical origin and socioprofessional category.