

# Epidemiology of sarcoidosis recorded in 2006–2010 in the Silesian voivodeship on the basis of routine medical reporting

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Kowalska M, Niewiadomska E, Zejda J.E. Epidemiology of sarcoidosis recorded in 2006–2010 in the Silesian voivodeship on the basis of routine medical reporting. *Ann Agric Environ Med.* 2014; 21(1): 55–58.

## Abstract

**Introduction and objective.** The incidence of sarcoidosis (D.86.0, D.86.2) varies worldwide, although published data suggest that the rate remains at the level 10–40/100 000 population. In Poland, statistics are not conducted on sarcoidosis. The etiology of the disease remains unclear, but researchers suggest that one of causes is the exposure to environmental factors. The aim of this study is to determine territorial and temporal variations of incidence and hospitalized prevalence of sarcoidosis for adults aged 19 and older living in the Silesian voivodeship in south-western Poland.

**Materials and methods.** To identify the number of cases of sarcoidosis and the number of hospitalized patients in 2006–2010, the database of the National Health Fund in Katowice, Silesia, was used. Data related to adults aged 19 and older who were inhabitants of the Silesian voivodeship. Standardized incidence and prevalence rates for total sarcoidosis per 100,000 population were calculated using the world standard population. Temporal and spatial variability of both rates in the Silesian voivodeship were presented according to ArcGIS 9.2.

**Results.** The number of new cases of sarcoidosis is still increasing. The disease occurred primarily in younger men (25–50 years of age), as well as in some older women (age group 50–64 years). Standardized incidence rates of total sarcoidosis are in the range 3.8–4.5/100,000 population. There was a probable relationship between the incidence of sarcoidosis and the area of forest ( $r=0.4$ ) or arable land ( $r=0.3$ ).

**Conclusions.** Sarcoidosis is a rare disease in the Silesian voivodeship and the standardized incidence and prevalence rates are slightly higher in men than in women. In the study period, spatial variability was observed. The highest rates were typical for districts with a predominance of forests and arable land.

## Keywords

Sarcoidosis, standardized incidence rate, standardized hospitalized prevalence rate, Poland

## INTRODUCTION

Sarcoidosis (D.86.0 and D.86.2 according to ICD-10) is an interstitial lung disease of unknown cause [1]. The incidence varies worldwide although published data suggest that the rate remains at the level 10 – 40/100,000 population [2]. Higher rates of incidence are noted in the population of Northern Europe, with a value 64/100,000 in Sweden and 43/100,000 in Germany. Smaller values are characteristic for the southern European countries, e.g., 9/100, 000 in Italy [3]. Current Polish data suggest that the incidence rate is in the rather low ranges (10/100,000) [3] and evaluated for people over 14 years old. In the Radom region of Poland, it was even smaller, at the level 2.3/100,000 population [2]. It is believed that this number may be underestimated because of the natural history of the disease and diagnostic problems [4]. Hospital mortality in patients with sarcoidosis is low and stands at 0.56%. It is worth noting that it is much smaller than in patients hospitalized for other interstitial lung diseases [2]. The disease usually affects young people, with a peak incidence between 20 – 29 years of age, and usually women over 50 years of age. The course of the disease varies in different populations, from a chronic lung disease to

disorders of calcium metabolism [3]. As already mentioned, the etiology of the disease remains unclear. The most likely cause of sarcoidosis is considered to be genetically-determined deficiency of cellular immunity [5, 6, 7, 8, 9, 10]. The results of the American ACCESS (A Case Control Study of Sarcoidosis Etiology) suggest a causal relationship between the disease and occupational exposure to insecticides and pollution mycobacteria [11]. These premises have become an argument for an epidemiological study plan aimed at fixing the recorded incidence and prevalence of hospitalized due to sarcoidosis in the Silesian region, and to assess their variability over time.

## OBJECTIVE

The aim of this study was to determine the territorial and temporal variation of incidence and hospitalized prevalence of sarcoidosis in adults aged 19 and older living in the Silesian voivodeship.

## MATERIAL AND METHODS

To identify the number of cases of sarcoidosis, and the number of patients treated in hospitals in the years 2006–2010, the database of the National Health Fund in Katowice, Silesia,

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Received: 25 February 2013; accepted: 28 September 2013

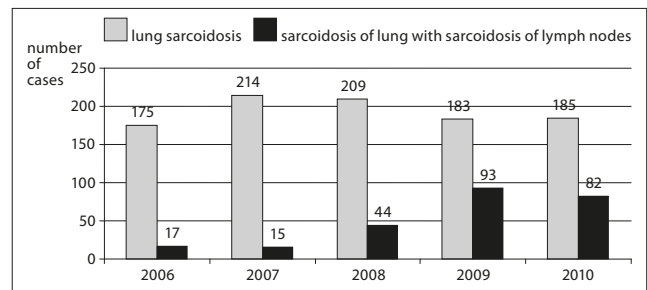
were used. It included the following information: metrical data, such as date of birth and gender of the patient, patient's administrative affiliation with regard to place of residence (to the nearest county), as well as data on the treatment, i.e., the main diagnosis, date of first illness, date of first hospitalization, number of further medical advice, number of subsequent hospitalizations, year of death. The analyses included only data of patients permanently domiciled in the Silesian voivodeship. Data related to adults aged 19 and older, were presented in a way that provide anonymity of patients. Sarcoidosis of lungs (D.86.0), and sarcoidosis of lungs with sarcoidosis of lymph nodes (D.86.2), were considered; both codes have been adopted by the International Classification of Diseases and Related Health Problems (ICD-10) [12]. For the presented study, the mean age of the patients was calculated, and the percentage of people hospitalized due to total sarcoidosis in 4 age groups: 19–34, 35–54, 55–64, and 65 or more years. The percentage changes during the study period was then examined, separately for men and women. Finally, crude prevalence and incidence rates for total sarcoidosis per 100,000 population were calculated. Crude rates were the direct standardized procedure using the WHO world standard population [13]. This assessment takes into account local differences and the territorial administrative division (NTS-4), including all counties and cities and towns in the province of Silesia. According to the definition, NUTS (Nomenclature of Territorial Units for Statistics) is an ordered list of names of political subdivisions and their associated symbols used in the collection of statistics by the Central Statistical Office (GUS) [14]. Maps were drawn showing variations for the period 2006–2010, averaged standardized values of incidence rates and hospitalization of total sarcoidosis in Silesian counties, using geographic information system ArcGIS 9.2 software.

## RESULTS

The results of basic analysis are summarized in Table 1. Data show that the number of new cases of sarcoidosis among adults aged 19 years and older in the Silesian voivodeship during the study period (years 2006–2010) is systematically growing. In addition, the increase of outpatient care participation in the diagnosis of sarcoidosis, which had been reported previously in the hospital, was observed. The highest number of deaths among people with diagnosed sarcoidosis (D.86.0 and D.86.2) was recorded in 2007. It is worth noting that in the Silesian voivodeship the most common form of

**Table 1.** Number of new cases, medical visits and number of hospitalization due to total sarcoidosis (D.86) in adults aged 19 years and older living in Silesian voivodeship

Year	Total cases N (100%)	No. of first-time medical visits N (%)	No. of first-time hospitalization N (%)	No. of medical outpatient visits N	No. of hospitalization N	No. of deaths N (%)
2006	192	1 (1%)	191 (99%)	7	323	8 (4.2%)
2007	229	3 (1%)	226 (99%)	5	371	21 (9.2%)
2008	253	10 (4%)	243 (96%)	50	422	6 (2.4%)
2009	276	36 (13%)	240 (87%)	75	404	6 (2.2%)
2010	267	53 (20%)	214 (80%)	75	267	7 (2.6%)



**Figure 1.** Number of registered incidence of lung sarcoidosis (D86.0) and sarcoidosis of lungs with sarcoidosis of lymph nodes (D86.2) in years in the Silesian voivodeship

the disease is sarcoidosis of lungs ( $n = 966$  out of all 1217 diagnoses in 2006–2010), which represents 79.4% of total cases (Fig. 1).

The average age of adult patients at onset in subsequent reporting years was 42.4 – 46.2 years. It is worth noting that the largest group of patients were those aged 35–54 years, and least numerous were older people, over 65 years of age (Tab. 2).

**Table 2.** Number of total cases of sarcoidosis (D.86) in adults aged 19 and older living in Silesian voivodeship by age

Year	Total population N	No. and percentage of cases				Average of the age $X \pm SD$
		19–34 years N (%)	35–54 years N (%)	55–64 years N (%)	65+ years N (%)	
2006	192	54 (28%)	95 (49%)	32 (17%)	11 (6%)	43.7±13.0
2007	229	61 (27%)	115 (50%)	35 (15%)	18 (8%)	44.6±12.6
2008	253	83 (33%)	118 (47%)	43 (17%)	9 (4%)	42.4±12.2
2009	276	54 (20%)	150 (54%)	48 (17%)	24 (9%)	46.2±12.6
2010	267	80 (30%)	110 (41%)	62 (23%)	15 (6%)	44.9±13.0

Figure 2 shows the changes in the age structure of people diagnosed with total sarcoidosis by gender in the following years. It should be noted that throughout the study period (2006 – 2010) medical doctors frequently recorded disease in younger men (between 25 – 50 years of age), as well as in some older women (age group 50–64 years).

According to the methodology, appropriate crude prevalence and incidence rates of total sarcoidosis for the Silesian voivodeship have been calculated, and next, both coefficients were standardized. (Tab. 3). Depending on the year, crude incidence rates of total sarcoidosis were in the

**Table 3.** Crude and standardized incidence and prevalence of total sarcoidosis (n/100 000) in general, male and female population aged 19 years and more, Silesian voivodeship

Year	Incidence (n/100 000 population) in adults aged 19 years and older				Prevalence (n/100 000 population) in adults aged 19 years and older			
	Crude rates	Standardized rates			Crude rates	Standardized rates		
		All	Women	Men		All	Women	Men
2006	5.1	3.8	3.3	4.3	5.1	3.3	3.2	3.2
2007	6.1	3.8	3.3	4.3	6.0	3.7	3.2	4.3
2008	6.7	4.3	3.4	5.2	6.5	4.1	3.2	5.0
2009	7.3	4.5	4.4	4.5	6.4	3.9	3.6	4.2
2010	7.1	4.3	3.5	5.0	5.7	3.4	2.8	4.1

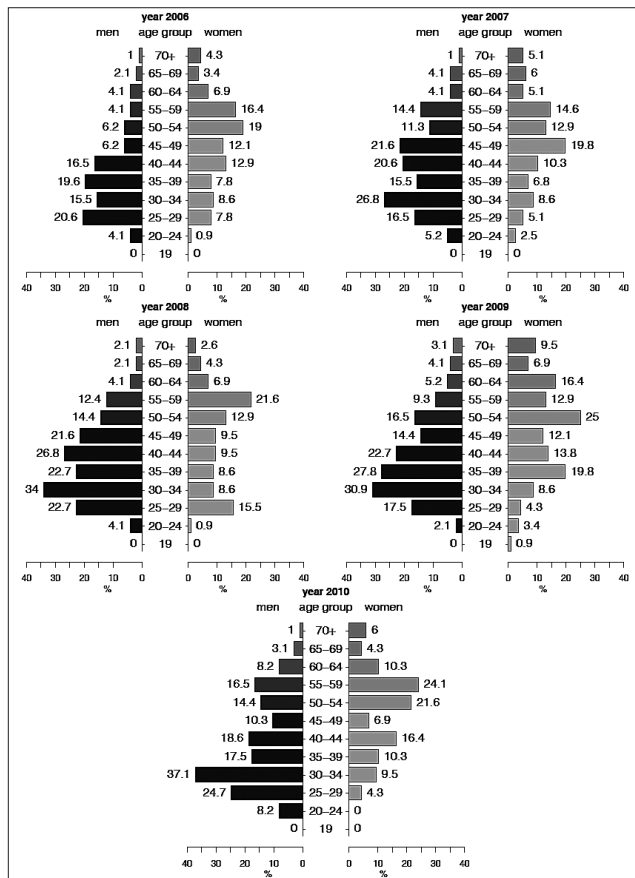


Figure 2. Percentage of new cases of total sarcoidosis (D.86) in adults aged 19 and older living in the Silesian voivodeship by gender and age in 2006–2010

range 5.1 – 7.3/100,000 population aged 19 and older, while the standardized rates were slightly smaller and amounted to 3.8 – 4.5/100,000.

Figure 3 shows the averaged values of standardized incidence rate of total sarcoidosis (the value was averaged for the period 2006–2010 in particular districts of the Silesian region), while Figure 4 reflects the diversity of territorial averaged values of hospitalized prevalence rate. In both cases, the highest values refers to Żywiec district and the cities of Częstochowa and Bielsko Biala.

DISCUSSION

According to the aim of the presented study, registry data on the incidence and hospitalized prevalence of sarcoidosis (D.86) in the Silesian voivodeship was analyzed. Among adults (persons aged 19 and over) who were residents of the region, from 192 – 276 cases of the disease per year were diagnosed. The predominant form of the disease is sarcoidosis of lungs (D.86.0 according to ICD-10), which accounts for over three-quarters of all diagnosed cases in the period 2006–2010. The standardized incidence rate of sarcoidosis was variable during the period and stood at 3.8 – 4.5/100,000 for the total population. It is worth noting that it was slightly lower in women (3.3 – 4.4/100,000) than men (4.3 – 5.2/100,000). These results suggest that Poland is a country with a rather low-incidence of sarcoidosis [2, 3].

Analysis of the data found that during the study period the disease was frequently recorded in younger men (25 – 34

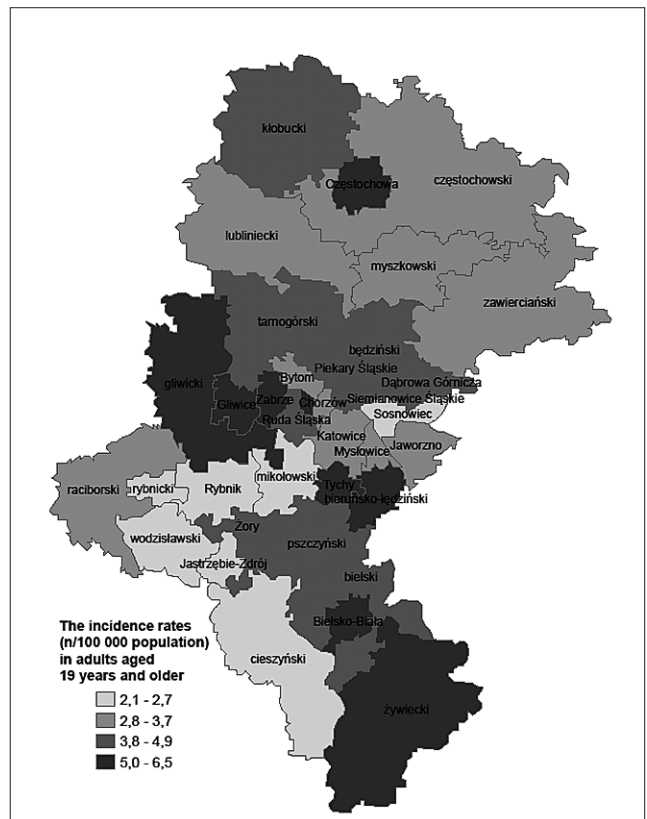


Figure 3. Mean value of the standardized incidence of total sarcoidosis in population of 19+ years (n/100 000), Silesian voivodeship, value averaged for the period 2006–2010

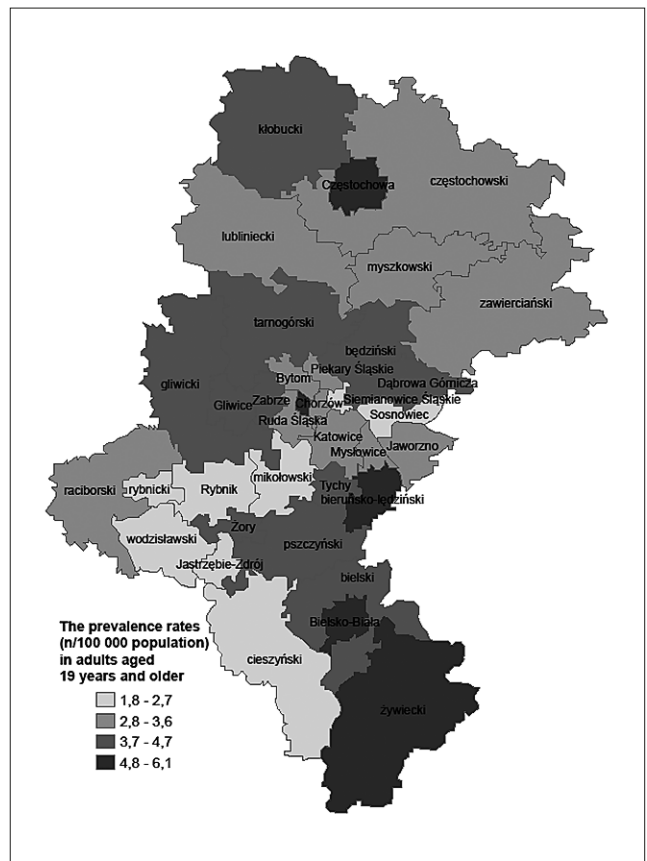


Figure 4. Mean value of standardized coefficient describes the number of first-time hospitalization for total sarcoidosis in a population of 19+ years (n/100 000) in the Silesian voivodeship. Value averaged for the period 2006–2010

years of age) and older women (50 years old). The age profile is consistent with previous observations by other authors [1, 3]. The percentage of patients with sarcoidosis who died were at the level 2.2–2.6% in the last years of observation (2008–2010). Unfortunately, the lack of information on the cause of death in the obtained database does not allow for the calculation of mortality. Available data indicate that specific mortality due to sarcoidosis is relatively low in Poland and does not exceed 0.6% [2].

There was considerable variation of averaged rate of incidences and hospitalization in the particular voivodships for the study period (years 2006–2010). The highest values of standardized incidence were recorded in Żywiec, Gliwice and Częstochowa counties. The highest prevalence occurred in Żywiec and Częstochowa counties. There is insufficient knowledge about the risk factors of sarcoidosis; therefore, this fact significantly impedes the explanation of the variation. Reports by other authors point to the potential role of exposure to environmental factors, such as occupational exposure to insecticides and mycobacteria [11]. From this perspective, it is worth noting that the largest incidence and hospitalization due to sarcoidosis occurred in districts with a majority of arable land and/or forest, where perhaps the use of plant protection products can also be larger [14]. Additional analyses showed a positive ecological correlation between the incidence of sarcoidosis and the area of forest ( $r = 0.4$ ) or arable land ( $r = 0.3$ ), in the absence of circumstances such as the importance – in districts – of the unemployment rate, average monthly salary, availability of medical doctors and hospital beds. The visualized ecological effect deserves verification during the study in order to follow the cause-effect relationships.

Among the other important reasons for epidemiological differentiation of sarcoidosis in the Silesian voivodeship is different access to diagnostic procedures. Against this type of interpretation, there are greater intensity ratios in districts with lower levels of urbanization. Distortion is also possible due to differences in the recording of cases of sarcoidosis. This issue can affect the observed territorial and temporal variation, but its outcome is not possible on the basis of available data.

It should also be noted that the hospitalized prevalence rates in the first years of observation (2006–2008) were similar to those of incidence. The last two years showed a marked reduction in the hospitalization rates of sarcoidosis, which may confirm the growing importance of outpatient care in the treatment of patients.

## CONCLUSIONS

In summarizing the obtained results, it is clear that sarcoidosis is a rare disease in the Silesian voivodeship, where the incidence is similar to that estimated for the Polish population. The standardized incidence and prevalence rates are slightly higher in men than women, and the proper variability in 2006–2010 was small. However, significant territorial differentiation was observed, with the highest value of both indicators in the districts of Żywiec, Gliwice, Częstochowa and Bielsko Białą.

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