# Prevalence of Behçet's disease in rural western Turkey: A preliminary report

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## ABSTRACT

**Objectives.** To study the prevalence of Behcet's disease (BD) in a rural area of Western Turkey.

Methods. This study was conducted in seven villages in the rural Havsa region which has a total population of 5727. The physicians and medical stu dents visited every household and dis tributed a questionnaire on the symp toms of rheumatic disorders. One of the questions was whether they had ever suffered from mouth ulcers. If the response was affirmative the proband was examined by a rheumatologist and a dermatologist for other symptoms. The individuals with true aphthous ulcerations had pathergy tests and were invited to the hospital for oph talmic examinations.

Results. Of the 5203 individuals who participated in this survey, 4861 were 10 years or older. 124 out of 190 indi viduals who responded positively to the question regarding the presence of mouth ulcers had true aphthous ulcera tions. 115/124 underwent a pathergy test and 4 showed positive reactions without any other stigmata of BD, ex cept for folliculitis in one. Among the individuals with a negative pathergy test, 2 had acneiform skin lesions and 1 a genital ulcer. No individual exhibited any ocular symptoms or findings cha racteristic for BD. Only 1 person with aphthous ulceration, folliculitis and a positive pathergy test was considered to have BD.

**Conclusion.** In this survey, the prevalence of BD was found to be 2: 10,000 (95% confidence interval 0 to 4.3: 10,000). This figure is lower than what has been reported in four previous studies from different regions in Turkey. This finding might be due to ethnic distribution and geographical differences.

## Introduction

Accurate estimates of disease prevalence are important. They are helpful in understanding disease pathogenesis, for diagnosis and in the distribution of health resources.

The prevalence of Behçet's disease (BD) shows considerable geographical variation. Reported rates show regional differences within a country and between countries. The condition seems to be most frequent in Mediterranean countries, the Middle East and Japan. Large epidemiological surveys are lacking and in many countries, prevalence figures are based on case registries and hospital attendance. The prevalence is about 13.5: 100,000 in Japan (1). and 0.64: 100,000 in England (2). In Turkey the prevalence rates were reported as 8 to 42: 10,000 in four previous surveys (3-6).

We conducted another field survey in a different region of Turkey where our clinical experience suggested the disease prevalence might be lower.

#### Subjects and methods

Seven villages in the rural Havsa region with a total population of 5,727 (according to government statistics) were included in the study. We report here our results which form part of a continuing larger study aimed at evaluating the prevalence of rheumatic diseases in the area. Havsa is a town in western Turkey (Thrace) on the border of Greece and Bulgaria. It was initially selected due to the clinical impressions of the local physicians who indicated that BD in this region could be uncommon in comparison to other parts of the country. The homogeneous socioeconomic status and proximity to a medical facility were the other factors in determining the study area. The physi-

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cians and specifically trained medical students visited every household and distributed a questionnaire on the symptoms of rheumatic disorders. One of the questions was whether the individuals surveyed had ever suffered from mouth ulcers. We used a positive answer to this question as the starting point of our survey.

If a proband replied positively to this question he/she was examined at the nearest local health care center by a dermatologist and rheumatologist for other symptoms of BD. The probands with true aphtous ulcerations had a pathergy test and were invited to the hospital for ophthalmologic examinations. The pathergy reaction is a non-specific hyper-reactive lesion which develops at the site of needle-prick in the patients with BD. The skin of the flexor aspects of the forearms was cleaned with disinfectants and pricked intradermally with a steril hypodermic needle (size 25G). Because of its specifity, especially at 48 hrs, the pathergy reaction was included in the diagnostic criteria proposed by the International Study Group for BD.

Since BD is quite rare in children under 10 years of age, this age group was excluded (7). The diagnosis of BD was made in accordance with The International Study Group for Behçet's Disease Criteria (8). According to this criteria set, recurrent oral ulceration (at least 3 episodes in one year) were taken as mandatory and the presence of at least two of the following findings was required for the diagnosis of BD: 1) genital ulceration; or 2) eye lesions (uveitis or retinal vasculitis); 3) skin lesions (erythema nodosum, papulopustular lesions, unexplained acne); and 4) a positive pathergy test.

The study protocol was approved by the hospital ethics committee.

### Results

The total population of the 7 villages surveyed was 5,727. A total of 5,203 individuals (mean age 38.4 yrs., M: F 0.99) responded to the questionnaire (93%) (Fig. 1). Excluding children less than 10 years of age, there were 4,861 individuals. Of those, 190 had a history of mouth ulcers and 178 of these agreed to be further evaluated. It turned out that 124/178 had true oral ulcers while the remainder had other oral lesions such cheliosis, denture trauma etc. The individuals with true oral ulcers were examined for other symptoms of BD (genital ulcers, erythema nodosum, acneiform lesions, folliculitis, and arthritis). 115 of the 124 individuals with true oral ulcers underwent pathergy tests. The remainder refused testing, stating either that they had no time or considered themselves healthy and did not wish to be included in this health survey. Those who underwent pathergy tests were also asked to come to the hospital for eye examinations. Pathergy was applied to the skin of the forearm at 6 different sites and an evaluation was made by the same physician after 48 hours. A papulo-erythema larger than 2 mm at any one of the sites was considered as positive. Pathergy was found to be positive in 4 individuals. Only one of these had folliculitis as a BD symptom. The others showed no symptoms. Of those individuals who had a negative pathergy test, two had acneiform lesions and one had a genital ulcer. The person with a genital ulcer underwent a repeated pathergy test and a monosodium urate (MSU) test. This test, described by us some time ago, more sensitive than the pathergy test, measures the skin response to intradermally injected monosodium urate crystals (9). Both results proved negative. None of the individuals exhibited any eye symptoms or signs compatible with BD.

At the end of this survey only one person with aphtous ulcerations, folliculitis and a positive pathergy test was considered to have BD. The MSU crystal test was also positive in this individual. In conclusion, BD was found to have a prevalence of 1/4861 (approximately 2:10,000, 95% confidence interval, 0 to 4.3:10,000) BD in this survey.

## Discussion

Four previous studies have focused on the frequency of BD in Turkey. The reported prevalence rates ranged from 8/10,000 to 42/10,000. The prevalence figures of BD were found to be 8: 10,000 in Silivri near Istanbul (3), 37:



Fig. 1. Results of the series design field study.

10,000 in the town of Camas in northern Anatolia (4), 11:10,000 in the Park region of Ankara (5), and 42:10,000 in a recent survey conducted in Istanbul (6).

In our study we estimated the prevalence of BD to be 2:10,000 in a rural population in Havsa.

There was no major difference between the method used in our study and those employed in previous studies and thus the differences noted were probably not due to methodological differences. Three of the areas studied (Camas, Silivri, Havsa) were rural. The other regions studied (Ankara, Istanbul) have sub-urban/urban characteristics. The distance between Istanbul, the region where the highest prevalence values were noted (6), and our area is not much greater than the distance between Silivri and our area. The distance from these two regions to our area is almost the same. Camas and Ankara are located quite far from Havsa. Istanbul is a large cosmopolitan city with a very high population motility. This phenomenon might contribute to increase the prevalence figures of BD as well as of other diseases. The populations of Camas and Ankara are made up of Anatolian Turks, while the population of Havsa is homogeneously made up of a higher proportion Balkan Turks. In brief, we consider that ethnic distribution represents an important factor in the lower prevalence we found in the current study.

Another aspect of the current study concerns the association of the frequency of oral ulcers in the general population with the prevalence of BD. Although oral ulceration is the most frequent symptom of BD, only 1 out of 115 individuals with oral ulceration had BD (8.7: 1000). In the Camas study (4) 19 out of 417 individuals with oral ulcerations had BD (45:1000) while in the Istanbul study (6) 101 out of 700 individuals with oral ulceration had BD (144:1000). Thus, neither the findings in the previous studies nor what has been observed in the current work seem to support the findings of Bang and colleagues from Korea, who reported that about 50% of individuals with oral ulceration go on to develop BD (10). This may again be due to ethnic, genetic, and geographical differences.

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