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Best Practice & Research Clinical Rheumatology

journal homepage: www.elsevierhealth.com/berh



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Behçet's syndrome and micro-organisms

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Keywords:

Behçet's syndrome
Micro-organisms
Infection
Epidemiology
Management

Behçet's syndrome (BS) is a multi-systemic vasculitis of unknown aetiology. More than one mechanism seems to be operative in the pathogenesis of Behçet's syndrome, including genetic and environmental factors, causing different manifestations of the syndrome. There are several clues to the role of environmental factors and especially micro-organisms in the pathogenesis. These include clinical findings such as a decrease in the frequency of a positive pathergy reaction with surgical cleaning of the skin before the procedure, the acne–arthritis association carrying similar features to acne-associated reactive arthritis, a higher rate of tonsillectomy, cold sores, late birth order, higher number of siblings, history of travel to countries with a high incidence of BS and earlier age at first sexual intercourse. Moreover, basic research on both viruses and bacteria suggests that micro-organisms may be playing a role, possibly through heat shock proteins and T-cell hypersensitivity.

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Introduction

Behçet's syndrome (BS) is a multi-systemic vasculitis involving the skin, mucosa, joints, eyes, veins and arteries of all sizes, gastrointestinal system and the central nervous system. There is evidence that more than one mechanism underlies the pathogenesis of BS, involving genetic and environmental factors, causing different manifestations of the syndrome. Here, we try to review data on the epidemiology, outcome and management of BS with special emphasis on the role of environmental factors, and especially micro-organisms in the pathogenesis of BS.

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Epidemiology

The classical comment on the geographic distribution of BS is that it is most prevalent along the ancient Silk Road, starting from countries around the Mediterranean Sea and the Middle East, extending to the Far East to Korea and Japan. This distinct geographical spread is referred to as an evidence for a genetic aetiology of BS [1]. However, it may also be indicating the role of environmental factors, especially infections that had spread along this famous route.

Prevalence of BS around the world

The reported prevalence of BS varies widely depending on the country, ethnic population and the methodology used. Even in Turkey, which is probably the country where BS is most prevalent, frequencies varying between 20 and 421 per 100 000 have been reported, depending on the area that is surveyed [2–6]. The first two field surveys reported from Turkey screened 4920 and 5131 people over the age of 10 according to O'Duffy's criteria, and found the prevalence as 80/100 000 and 370/100 000 [2,3]. Human leucocyte antigen (HLA) B51 was positive in 75% and 26% of patients, respectively, but in the first study there were only four patients with BS. The next two studies were done in big cities, Ankara and Istanbul, using International Study Group (ISG) criteria [4,5]. The prevalence was 115/100 000 in the Ankara study. In the study from Istanbul, people over the age of 12 were screened and the prevalence was 421/100 000, the highest prevalence ever reported for BS. In a more recent field survey from the western part of Turkey, the prevalence was 20/100 000 [6] and another study among children surveyed 46 813 children and none of them had BS [7].

Studies from countries in the Far East, Middle East and North Africa report somewhat lower prevalences. A nationwide hospital-based survey of BS in Japan showed an estimated prevalence of 11.9/100 000 [8]. There is an impression that the prevalence of BS is decreasing in Japan. In a report from Hokkaido University, the number of BS patients that presented to their clinic between 1999 and 2002 was 7.2 per year compared to 30 per year between 1978 and 1983 [9]. They also reported that the severity of uveitis had decreased over the years [10]. Field surveys from Iraq showed a prevalence of 17/100 000 [11], from Iran 80/100 000 [12] and from Saudi Arabia 20/100 000 [13]. In Egypt, the prevalence was 7.6/100 000 [14]. There are two reports on Arabs living in Israel [15,16]. In one of them, the prevalence is 120/100 000 [15] and in the second it is reported as 26/100 000 [16]. In the same survey, prevalence of BS was 146/100 000 among Druises living in Israel and 8.6/100 000 among Jews, giving an overall prevalence of 15.2/100 000.

In Europe, the prevalence shows a general decrease as one goes from the South to the North. In Italy, Portugal, Spain and among French living in Paris, prevalences are relatively high, between 1.5 and 7.5/100 000, compared to Scotland, UK, Sweden and the Germans in Germany (0.3, 0.64, 1.2 and 1.47 per 100 000, respectively) [17–25].

In a retrospective population-based study from Olmsted County, Minnesota, US, the prevalence was reported as 5.2/100 000 and the incidence was 0.38/100 000 [26].

Clues from epidemiology for genetic versus environmental aetiology

Determining the prevalence of a disease in ethnic populations, who have migrated to parts of the world other than from which they have originated, and comparing it to that in the original area is considered as a good way of determining whether genetic or environmental factors have more influence on the aetiology. There are several examples to such studies in BS. In Berlin, the prevalence of BS among people of Turkish origin was 77.37/100 000 [25]. This is a much higher prevalence compared to the Germans in the same study (1.47/100 000), but somewhat lower than the prevalence reported by most studies reported from Turkey. A survey of BS among Armenians over the age of 10, living in Istanbul, showed a prevalence of 110/10 000 [27], whereas, as mentioned above, the prevalence of BS in a field survey in Istanbul was 421/100 000. Another interesting study surveyed the prevalence of BS in Paris among European, North African, Asian and Turkish, sub-Saharan African and non-continental French populations by capture–recapture method [21]. The prevalence of BS among these populations varied from 2.4/100 000 among Europeans to 17.5 among Asians including Turks and 34.6

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