CONCISE REPORT

Prevalence of Behçet's disease in an Arab community in Israel

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Objective: To evaluate the prevalence of Behçet's disease (BD) in an Israeli Arab town (Taibe).

Methods: Questionnaires about the occurrence and prevalence of aphthous ulcers were distributed randomly to the parents of children attending a paediatric centre in Taibe. The parents were asked whether they or any of their children aged between 10 and 20 years had recurrent aphthous stomatitis. Any who had had more than four aphthous episodes (each episode lasting more than seven days) during the previous year were invited for an extensive interview and examination by a rheumatologist or a paediatrician.

Results: A total of 4876 subjects were included in this survey, of whom six (one male, five female) were diagnosed as having BD. Of these six, two were siblings (a brother and a sister). Five had skin lesions, four had visual involvement, and all had genital ulcers and joint symptoms; one in two patients had a positive pathergy test. Five of the six carried HLA-B5 antigens. The results showed a prevalence of 12/10 000 in Taibe.

Conclusion: The prevalence of BD found in our survey is high and concurs with that found in other Mediterranean and Asian countries.

Behçet's disease (BD) is a chronic multisystem disorder of unknown cause. The main features are recurrent aphthous stomatitis (RAS), genital ulcerations, and ocular disease, as well as mucocutaneous, articular, neurological, urogenital, vascular, intestinal, and pulmonary manifestations.¹ Although BD occurs world wide, it is generally regarded as being most common in the Mediterranean basin, the Middle East, and the Far East. The prevalence of BD varies in different parts of the world. A field survey in Turkey found a prevalence of 8/10 000 in nine villages near Istanbul,² and a higher prevalence in a rural area in northern Turkey.³ On the other hand, in Hawaii, a study carried out by a questionnaire/letter to determine the number of patients with BD showed a prevalence of zero,⁴ whereas a similar study in Japan found a prevalence of 7–8.5/100 000.⁵

POPULATION AND METHODS

We conducted a field survey to evaluate the prevalence of this condition in Taibe, a large Arab town in the centre of Israel, founded 200–300 years ago by 10–12 original families, and which has a population of about 30 000. Questionnaires were randomly distributed on a first come basis to 2211 fathers or mothers of children attending the Bridge to Peace Community Paediatric Centre in Taibe, which provides health care to approximately 7000 children from birth to 15 years of age. The parents were asked whether they or any of their children, including those who did not attend the clinic, between the ages of 10 and 20 years had RAS. The questionnaires were

filled out by the doctors who interviewed the parents. Any parent or child with more than four episodes of aphthous stomatitis (each episode lasting more than seven days) during the previous year was invited for an extensive interview and examination. During the interview, the parents were questioned and they and the children were examined for the presence of genital ulceration, dermal vasculitis, thrombophlebitis, folliculitis or erythema nodosum-like lesions, arthritis/arthritis, uveitis or central nervous system (CNS) involvement.

RESULTS

Of the 2211 questionnaires distributed, 1938 (87.7%) were returned, which reported on a total of 4876 subjects—1888 adults and 2988 children. A total of 1609 reported recurrent mouth lesions, of whom 360 were children. Seven hundred and sixty of the 1609 subjects were excluded because it was considered that their mouth lesions represented problems other than aphthous stomatitis. Eight hundred and forty nine people were diagnosed as having true RAS, and of these, 829 (685 adults, 144 children) underwent physical examination. Among these, six adults (one male, five female) were found to fulfil the criteria for Behçet's disease according to the International Study Group for Behçet's disease⁶ (table 1).

DISCUSSION

Our study shows that in the Arab town of Taibe in Israel, the prevalence of BD among the population aged 10 years and over is approximately 12/10 000. Our results are consistent with those found in a Turkish population (8/10 000),² higher than those found in the Japanese population (7–8.5/100 000),⁵ and vary considerably from those found in another Turkish study, which was carried out in a different location and which showed a prevalence of 37/10 000.³ The prevalence of BD in our study is much higher than that reported for America and Europe combined, which is 0.1–7.5/100 000.⁷ It therefore appears that the prevalence of BD varies both between countries and between different regions of the same country. These regional differences are probably due to ethnic as well as geographical factors, and may be a result of the founder effect.^{2,3} Indeed, each Arab village in Israel was founded by a small number of families who intermarried and expanded. Within the Israeli Arab community 44% of all marriages are consanguineous,⁸ with more than half of these being between first cousins, leading to several genetic diseases and congenital malformations.

Our previous studies in the Arabic community in Israel,⁹ as well as studies from other countries, disclosed a male to female ratio of BD of 1.2:1 to 3.4:1.^{10–13} The present ratio of 1:5 might be due in part to the fact that mostly mothers attended the clinic.

Abbreviations: BD, Behçet's disease; CNS, central nervous system; RAS, recurrent aphthous stomatitis

Table 1 Disease expression (%) of BD in Arabs¹⁰⁻¹³

	Kuwait	Jordan	Saudi Arabia	Iraq	Israeli Arabs	Present study
Number of patients	29	20	119	60	28	6
Ratio of men/women	3.1	2.3	3.4	3	1.2	0.2
Aphthous stomatitis	100	100	100	97	100	100
Genital ulcers	93	65	87	83	79	100
Ocular symptoms	69	65	85	48	56	66.7
Skin involvement	76	25	57	48	68	66.7
Joint manifestations	69	55	37	48	41	100
Neuro-Behçet	21	5	44	0	15	16.6
GI involvement	21	10	4	0	14	0
Pulmonary system	7	5	16	0	0	0
Vascular system	34	45	40	17	26.5	16.6
Pathergy test	34	20	17.5	71	36.8	50.0
Family history	Not reported	25	Not reported	Not reported	Not reported	33
HLA-B5	Not reported	Not reported	72	62	76.2	83

Our results of the clinical expression of BD as well as HLA-B51 association are in line with reports from Arab countries such as Saudi Arabia, Iraq, Jordan, and Kuwait¹⁰⁻¹³ insofar as the percentage of patients with genital ulcers, skin manifestations, and CNS involvement in our study is similar to that found in most of the series studied in those countries. Forty four per cent of marriages in the Arab community are consanguineous. A positive family history was noted in two of our six patients with BD. Most cases of BD are sporadic and the parents of the patients are unaffected, but a familial aggregation has been previously reported.¹⁴ A high sibling recurrence risk ratio for BD indicates that strong genetic factors play a part in the mechanism for the familial incidence in this syndrome.

In conclusion, the prevalence of BD found in our survey is high, it concurs with that found in other Mediterranean and Asian countries, and might be attributed, in part, to a founder effect compounded by the high incidence of consanguineous marriages in this population.

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