Letters to the editor

Prevalence of Behçet's disease in the province of Brescia in northern Italy

Sirs,

Studies calculating the prevalence of Behçet's disease (BD) in Italy have reported considerable variation across latitude (1-3), ranging from 3.8/100.000 (95%CI 2.0–5.8) in Reggio Emilia, northern Italy, to 15.9 (8.9–28.5) in Potenza, southern Italy. In these studies, patients were classified as having BD if they met the International Study Group (ISG) criteria (4), but new International Classification Criteria (ICBD) with improved sensitivity have subsequently been developed (5); only patients of Italian ancestry were identified, but this result might be influenced by the low presence of immigrants in Potenza (3), and the relatively low number of patients recruited.

A further evaluation of BD prevalence in Italy might be useful for several reasons: to confirm the differences between northern and southern regions of the country, to evaluate the prevalence in an area with a high immigration rate, and to evaluate the impact of ICBD classification. For these purposes, we have evaluated the prevalence of BD in the province of Brescia, an area of northern Italy, in which a large number of residents are immigrants.

Patients were recruited from two sources: the electronic databases of our Unit and the regional registry for rare diseases. In all cases, the clinical diagnosis of BD was reassessed by a senior rheumatologist, and both ISG criteria and ICBD were applied to classify patients.

As January, 2012, 50 patients (28 male; 22 female) with a confirmed clinical diagnosis of BD lived in our province (population: 1,056,063 inhabitants older than 14 years; immigrants; 129,296. Data source: ISTAT); 11 were first-generation immigrants of non-Italian origin (Morocco: 5; Albania: 3; Egypt: 2; Senegal: 1); 43 of them met the ISG criteria, and 46 the ICBD. The estimates for the point prevalence are reported in Table I.

Table I. Prevalence of BD per 100,000 inhabitants older than 14 years in province of Brescia, categorised by sex and age. The 95% confidence intervals were calculated according to the Poisson distribution.

	Total population (n=1,056,063)	Italian-origin population (n=926,767)	Non Italian-origin population (n=129,296)
Prevalence (ISG)	4.1 (C.I.: 2.9-5.2)	3.5 (C.I: 2.3-4.6)	8.5 (C.I: 3.5.-13.5)
Prevalence in males (ISG)	4.9 (C.I.: 3.0-6.8)	3.4 (C.I.: 1.7-5.2)	14.8 (C.I: 5.7-23.9)
Prevalence in females (ISG)	3.3 (C.I: 1.8-4.9)	3.5 (C.I: 1.8-5.2)	1.6 (C.I: 0-4.8)
Prevalence (ICBD)	4.4 (C.I.: 3.1-5.6)	3.8 (C.I: 2.5-5.0)	8.5 (C.I: 3.5.-13.5)
Prevalence in males (ICBD)	4.9 (C.I.: 3.0-6.8)	3.4 (C.I.: 1.7-5.2)	14.8 (C.I: 5.7-23.9)
Prevalence in females (ICBD)	3.9 (C.I: 2.2-5.5)	4.2 (C.I: 2.4-6.0)	1.6 (C.I: 0-4.8)
Prevalence (clinical diagnosis)	4.7 (C.I.: 3.4-6.0)	4.7 (C.I.: 3.4-6.0)	8.5 (C.I: 3.5.-13.5)
Prevalence in males (clinical diagnosis)	5.4 (C.I.: 3.4-7.4)	4.0 (C.I: 2.0-5.9)	14.8 (C.I: 5.7-23.9)
Prevalence in females (clinical diagnosis)	4.1 (C.I: 2.4-5.8)	4.4 (C.I: 2.5-6.3)	1.6 (C.I: 0-4.8)

The results allow the following conclusions: first, the prevalence of BD in our province is similar to that reported in another area of northern Italy, and lower than in southern Italy. Second, among males, the crude prevalence in immigrants is higher than in Italians, similarly to what was observed in France evaluating the ethnic origin of BD patients (6). Morocco and Egypt are estimated to be among the countries with the highest BD prevalence after Turkey (7-10). No data on BD prevalence in Albania are available, but this country was under the Turkish rule for five centuries, and has a rather high prevalence of HLA-B51, a risk factor for BD (10). In keeping with the literature, BD is more prevalent in males than in females among immigrants, whereas it is equally frequent in Italians (1-3,11). Third, the use of the ICBD slightly increases the prevalence of BD, in accordance with the 10% increase of sensitivity, as compared with the ISG criteria, calculated by the team who developed the ICBD (5).

Our hospital is the only one in the province allowed to certificate the diagnosis of BD in the regional rare diseases registry (certification is mandatory in order for the patient to be exempt from payment of disease-related medical costs), and other specialists (*e.g.* dermatologists, ophtalmologists, etc.) refer patients with BD to our Unit for systemic therapy. Nevertheless, we cannot exclude that patients with milder forms of BD, not requiring systemic therapy or certification, were missed. Indeed, we have observed a more severe disease (a higher frequency of permanent ocular or neurological damage) in immigrants than in Italians (12). Although this might be explained by a milder course of disease in Italians (1), we cannot exclude that, among immigrants, only patients with more severe disease seek medical evaluations. This might lead to an underestimation of the prevalence of BD among these populations. Late referral might be another cause of more severe disease course in immigrants.

S. CARTELLA, *MD*
M. FILIPPINI, *MD*
A. TINCANI, *MD*
P. AIRÒ, *MD*

Rheumatology and Clinical Immunology Unit, Spedali Civili and University of Brescia, Italy;

Address correspondence and reprint requests to: Paolo Airò, Rheumatology and Clinical Immunology Unit, Spedali Civili of Brescia, P.le Spedali Civili 1, 25123 Brescia, Italy. E-mail: airo@bresciareumatologia.it

References

1. SALVARANI C, PIPITONE N, CATANOSO MG *et al*.: Epidemiology and clinical course of Behçet's disease in the Reggio Emilia Area of Northern Italy: a seventeen-year population-based study. *Arthritis Rheum* 2007; 57: 171-8.
2. LECCESE P, D'ANGELO S, GILIO M *et al*.: Prevalence of Behçet's disease in an urban area of south of Italy (abstract). *In*: 48. Congresso Nazionale della Società Italiana di Reumatologia. *Reumatismo* 2011; 63; S3:110.
3. OLIVIERI I, LECCESE P, PADULA A *et al*.: High prevalence of Behçet's disease in southern Italy. *Clin Exp Rheumatol* 2013; 31 (Suppl.77): S28-31.
4. INTERNATIONAL STUDY GROUP FOR BEHÇET'S DISEASE: Criteria for diagnosis of Behçet's disease. *Lancet* 1990; 335: 1078-80.
5. INTERNATIONAL TEAM FOR THE REVISION OF THE INTERNATIONAL CRITERIA FOR BEHÇET'S DISEASE (ITR-ICBD): The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. *J Eur Acad Dermatol Venereol* 2014; 28: 338-47.
6. MAHR A, BELARBI L, WECHSLER B *et al*.: Population-based prevalence study of Behçet's disease. Differences by ethnic origin and low variation by age at immigration. *Arthritis Rheum* 2008; 58: 3951-9.
7. MAHR A, MALDINI C: Épidémiologie de la maladie de Behçet. *Rev Med Interne* 2014; 35: 81-9.
8. BENAMOUR S, ZEROUAL B, BENNIS R, AMRAOUI A, BETTAL S: Behçet's disease. 316 cases. *Presse Med* 1990; 19: 1485-9.
9. ASSAAD-KHALIL S, KAMEL F, ISMAIL E: Starting a regional registry for patients with Behçet's disease in North West Nile Delta region in Egypt. In: HAMZA M. (Ed.). *Behçet's Disease*. Tunis (Tunisia): Pub Adhoua; 1997: 173–6.
10. VERITY DH, MARR JE, OHNO S, WALLACE GR, STANFORD MR: Behçet's disease, the Silk Road and HLA-B51: historical and geographical perspectives. *Tissue Antigens* 1999; 54: 213-20.
11. TALARICO R, D'ASCANIO A, FIGUS M *et al*.: Behçet's disease: features of neurological involvement in a dedicated centre in Italy. *Clin Exp Rheumatol* 2012; 30 (Suppl. 72): S69-72.
12. CARTELLA S, FILIPPPINI F, FRASSI M, FRANCESCHINI F, AIRO' P, TINCANI A: Manifestazioni cliniche della sindrome di Behçet in 80 pazienti di eterogenea provenienza geografica (abstract). *In*: 50. Congresso Nazionale della Società Italiana di Reumatologia. *Reumatismo* 2013; 65; S3: 509.