

Prevalence of Adamantiades- Behçet's disease in Germany and the municipality of Berlin: Results of a nationwide survey

Sirs,

The worldwide prevalence of Adamantiades-
Behçet's disease (ABD) has been esti-
mated to be in the range of 0.12 - 420 per
100.000 inhabitants, the highest assessed in
Istanbul, Turkey (1) and the lowest in the
U.S.A. (2). This considerable difference is
likely to be genetically determined as fur-
ther suggested by occasional familial cases
and in association with HLA haplotypes,
such as HLA-B₅₁; though it remains unclear
whether exogenous agents trigger the dis-
ease (3).

The current prevalence of ABD in Germany
based on data of the German registry that
has been established in 1990 was higher
than previous reports not only among native
Germans but also among other ethnic
groups. The 2005 survey estimated a preva-
lence of ABD in Germany of 0.72: 100.000
and in the municipality of Berlin of 4.87:
100.000. This current increase, however, is
still in the range of ABD prevalence in
other North- and West-European countries
(4, 5). From the registered 590 patients (344
male and 246 female), 522 patients fulfilled
the criteria of ABD classification tree (6),
63 additional patients fulfilled Cheng and
Zhang's criteria (4 complete and 59 incom-
plete) (7), one additional patient fulfilled
Dilsen's criteria (8), and 4 patients were
diagnosed clinically in the initial stage of
the disease only presenting recurrent oral
aphthae. The patients belonged to 31 ethnic
groups: 267 were of Turkish origin, fol-
lowed by patients of German (n = 227),
Greek (n = 10), Italian (n = 10), Lebanese
(n = 10), Serbian and Montenegrin (n = 9),
Vietnamese (n = 5), Moroccan (n = 5) and
other, less frequent origin (n = 49). Of the
165 ABD patients registered in the munic-
ipality of Berlin, the patients of Turkish ori-
gin constituted the larger ethnic group (n =
91), followed by patients of German (n =
43), Lebanese (n = 8), Greek (n = 4), Italian
(n = 3) and other origin (n = 16).
The current nationwide prevalence of ABD

among the German population is 0.30 per
100.000, whereas in Berlin it is 1.47:
100.000. The highest ethnic prevalence was
that of patients of Turkish origin (nation-
wide 15.13: 100.000; in Berlin 77.37:
100.000), as well as by patients of Lebanese
(nationwide 24.46: 100.000; in Berlin
101.32:100.000) and Greek origin (nation-
wide 3.16: 100.000; in Berlin 39.73:
100.000). Previous reports of ABD preva-
lence in Berlin, the city with the highest
number of non-native German inhabitants,
have shown lower rates (Table I). Al-
though there was an increase of ABD
prevalence in all ethnic groups, the most
striking increase is among non-native Ger-
mans, mainly among Turks. Interestingly,
the current prevalence in Berlin among
patients of Turkish origin has approached
that of Western Turkey (20-80: 100.000)
(9). It is likely that this increase is not only
due to new cases from all ethnic groups but
also to increased awareness for the disease.
Furthermore, most of the ABD patients eth-
nic groups (Turks, Lebanese, Kurds, Kaza-
khs, Kyrgyz, Iranians, Afghans, Indians,
Pakistanis, etc.) represent those living along
the historical "silk road" thus favouring the
genetic aetiology of the disease rather than
environmental factors (10).

The patients of Turkish origin exhibited an
androtropism with a male-to-female ratio of
1.9 (175 males and 92 females), whereas
male patients only were registered in a few
other ethnic groups from Southern Euro-
pean and the Middle East. German patients
exhibited an almost equal male-to-female
ratio (119 females and 108 males); ethnic
groups of Far East, such as Koreans and
Vietnamese only included female ABD
patients. Obviously the historical ABD
androtropism seems to vanish as surveys
become larger and carried out nationwide.
Indeed, in surveys with large numbers of
ABD patients, no gender prevalence was
assessed.

In conclusion, the prevalence of ABD in
Germany calculated in this study is higher
than in earlier reports but it still ranks
among the prevalence in other north-west-
ern European populations. The prevalence
of ABD in Berlin is the highest nationwide,
due to the multi-ethnic character of the

urban Berlin population. The observed pre-
valence in patients of Turkish origin living
in Germany was similar to that reported
from Western Turkey. This finding disputes
previous concepts of an environmental trig-
gering factor and corroborates the major
role of a genetic background in ABD patho-
genesis.

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Table I. Development of prevalence in Berlin.

Population	1984		1989		1994		2005	
	pat. / pop.	Pro	pat. / pop	Pro	pat. / pop.	Pro	pat. / pop.	Pro
All patients	13/2147943	061	35/2202734	159	44/2170411	202	165/3391344	487
Female	5/1144656	044	21/1154059	182	25/1135762	220	77/1734495	444
Male	8/1003317	080	14/1048675	134	19/1034649	184	88/1656849	531
German	3/1903856	016	8/1915405	042	10/1817211	055	43/2932755	147
Non-German	10/244117	410	27/287329	940	34/353200	963	122/458589	2660
Turkish			24/125297	1915			91/117624	7737