Insights into Adamantiades-Behçet Disease in Germany: A Focus on Prevalence, Treatment Patterns, and Clinical Characteristics

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Introduction

In many European countries Adamantiades-Behçet Disease (ABD) poses diagnostic and therapeutic challenges due to its rarity and varied clinical presentations. This study aims to analyze ABD in Germany, focusing on prevalence, incidence, clinical manifestations, and treatment. ABD is a rare disease, with a reported global prevalence of 10.3/100,000 population (95% confidence interval [CI]: 6.1,17.7). There is substantial geographic variation in its prevalence, which is higher along the Silk Road, an ancient trading route (Turkey: 119.8/100,000; Middle East: 31.8/100,000), compared with other regions (Asia: 4.5/100,000; Europe: 3.3/100,000).

Method

Data from the German Registry of ABD (>900 patients) were compared with preliminary data from the Institute for Applied Health Research Berlin database (see Ref.). To determine the final analysis population for prevalence and incidence, a step-wise approach to patient selection was undertaken: To evaluate prevalence, ≥2 confirmed outpatient diagnoses in different quarters (minimum 2 quarters) or ≥1 main inpatient diagnosis of ABD in the observation year were required; secondary inpatient diagnoses were treated as outpatient diagnoses Prevalence, incidence, and treatment patterns were evaluated.

Table 1. Prevalence of ABD in patients receiving a prescription for disease-related medication between 2016 and 2018 (SHI, statutory health insurance)

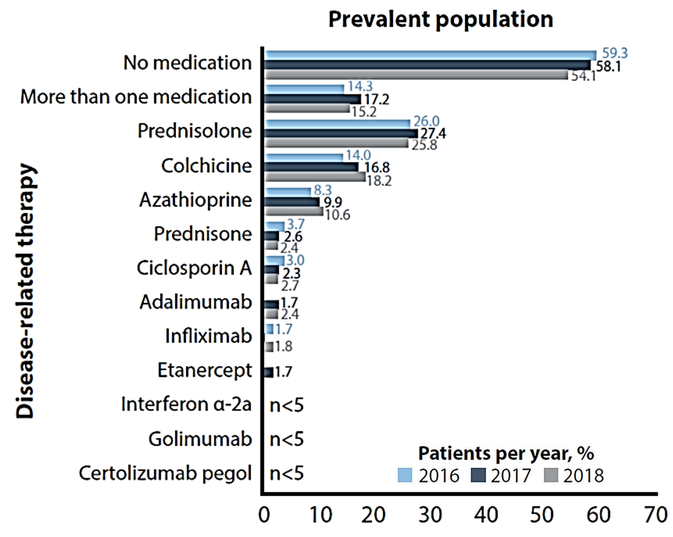
	2016	2017	2018
Prevalent patients			
N (M/F)	122 (62/60)	127 (64/63)	150 (79/71)
Mean (SD) age, years			
Overall	50.1 (13.7)	50.7 (14.3)	51.7 (13.7)
Male	48.1 (13.4)	49.4 (13.6)	50.7 (12.8)
Female	52.1 (13.8)	52.0 (15.0)	52.8 (14.6)
Prevalence (95% CI), adjusted per 100,000 population			
Overall	3.9 (3.2, 5.8)	4.1 (3.4, 5.9)	4.7 (4.0, 6.4)
Male	4.2 (3.0, 6.2)	4.2 (3.2, 6.4)	5.3 (3.9, 7.3)
Female	3.8 (2.9, 7.3)	3.9 (3.0, 7.2)	4.4 (3.4, 7.5)
M:F ratio	1.03	1.02	1.11
Prevalence (95% CI), adjusted per 100,000 population, by age group			
18–39 years	4.5 (3.1, 7.8)	4.2 (2.8, 7.4)	3.6 (2.3, 6.7)
40–59 years	4.8 (3.5, 7.1)	5.3 (4.0, 7.6)	7.4 (5.8, 10.0)
60+ years	2.6 (1.7, 7.7)	2.7 (1.8, 7.5)	3.1 (2.2, 7.5)

CI, confidence interval; N, total number of patients; SD, standard deviation

Discussion

ABD typically manifests in the third decade, with oral aphthae as the primary symptom. Patients of Turkish descent showed higher rates of positive familial history and uveitis prevalence. HLA-B5 positivity correlated with certain clinical manifestations, including uveitis and gastrointestinal involvement. A cohort study indicated a steady rise in ABD prevalence (estimated prevalence in Germany approximately 4.2 : 100,000). Prednisolone, colchicine, and azathioprine were common treatments, with 15% receiving combination therapy. In the treated population, ABD prevalence was 3.9 (2016), 4.1 (2017) and 4.7 (2018) per 100,000 population (Table 1); annual ABD incidence was 0.5 per 100,000 population in 2016 and 2017 and 0.6 per 100,000 population in 2018.
The most commonly reported comorbidities in patients diagnosed with ABD were dorsalgia, been an indicator of possible misdiagnsosis, disorders of refraction and accommodation, and essential (primary) hypertension. Prednisolone, colchicine and azathioprine were the most commonly prescribed treatments for ABD, with approximately 15% of patients taking >1 medication for ABD.

Figure 1: Disease-related medications between 2016 and 2018 in patients with ABD in the prevalent population



Conclusion

In Germany, recent cohort studies have indicated a notable rise in ABD prevalence, aligning with the trend in our registry. The estimated prevalence of approximately 4.2 cases per 100,000 underscores its growing recognition in the healthcare system

Source database / References

Source database: German Registry of Adamantiades-Behçet Disease References: Zouboulis, C.C., et al. (2021). Prevalence and Incidence of Adamantiades-Behçet's Disease: An Epidemiological Study from Germany. In: German Rheumatology Congress 2021, 49th Congress of the German Society for Rheumatology (DGRh), September 15th - 18th, 2021, virtual. [URL: https://www.egms.de/static/en/meetings/dgrh2021/21dgrh057.shtml]
Zouboulis CC, Kotter I, Djawari D, Kirch W, Kohl PK, Ochsendorf FR, et al. Epidemiological features of Adamantiades-Behcet's disease in Germany and in Europe. Yonsei Med J. 1997;38:411–22.
Zouboulis CC. Epidemiology of Adamantiades-Behcet's disease. Ann Med Interne (Paris). 1999;150:488–98.
Maldini C, Druce K, Basu N, LaValley MP, Mahr A. Exploring the variability in Behcet's disease prevalence: a meta-analytical approach. Rheumatology (Oxford). 2018;57:185–95.