TABLE 1.

	Diagnostic criteria and study period	n cases / population size	Mean age at onset and sex (%female)	Prevalence (over 100000) / incidence	Oral / genital ulcers (%)	Skin lesions/ pathology test (%)	Ocular involvement (%)	Joint involvement (%)	Neurobehcet/ Vascular/ Gastrointestinal involvement (%)
Sánchez, L. et al. Southern Europe (Cantabria, Spain)	Expert opinion, ISG, ICBD / 1980- 2020	120 (expert opinion) / 96 (ICBD) / 59 (ISG) / 581641	37.6 ±13.8/ 48.3	20.6 (expert opinion), 16.5 (ICBD), 10.1 (ISG) / 0.022 (expert opinion), 0.018 (ICBD), 0.011 (ISG)	94.2 / 59.2	63.3 / 25.2	41.6	65	10.8 / 11.6 / 6.6
Calamia, K. T. et al. North America (Minnesota, USA)	ISG / 1960-2005	13 / NR	31 / 30	5.2 / 0.38	100 / 62	85 / NR	62	46	23 / 23 / NR
Altenburg, A. et al. Northern Europe (Berlin, Germany)	ISG and ABD classification tree / 1961-2005	590 / 3391344	26 / 58	4.9 / 1 (estimated)	98.5 / 63.7	62.5 / 33.7	58.1	53	10.9 / 22.7 / 11.6
Mohammad, A. et al. Northern Europe (Skane County, Sweden)	ISG / 1997-2010	40 / 809317	30.5 / 33	4.9 / 0.2	100 / 80	88 / NR	53	40	0 / 20 / NR
Mahr, A. et al. Southern Europe (SeineSaint-Denis County, France)	ISG / 2003	79 / 1094412	27.6 / 43	7.1 / NR	100 / 80	90 / 20	51	59	10 / NR / 10
Salvarani, C. et al. Southern Europe (Reggio Emilia, Italy)	ISG, 1988-2005	18 / 486961	33 / 50	3.7 / 0.24	100 / 78	100 / NR	56	50	11 / 6 / NR
Azizlerli, G. et al. Middle East (Istanbul, Turkey)	ISG / prevalence study	101 / 23986	NR / 48.5	42 / NR	100 / 70.2	Not globally reported / 69.3	27.7	Not globally reported	NR / Not globally reported / NR
Davatchi, F. et al. Middle East (Iran nationwide)	Expert opinion / 1975-2018	7641 / NR	25.6 / 44.2	80 / NR	97.5 / 64.4	62.2 / 50.4	55.6	38.1	3.9 / 8.9 / 6.8
Krause, I. et al. Middle East (Galilee, Israel)	ISG / 15 years (not specific years have been reported)	112 / 737000	30.6 / 47	15.2 / NR	NR / 68	41 / 44.4	58	70	11.6 / Not globally reported / NR
Nishiyama, M. et al. Asia (Japan nationwide)	1987 JCBD / 1991	3316 / NR	35.7 / 50.6	NR / NR	98.2 / 73.2	87.1 / 43.8	69.1	56.9	11 / 8.9 / 15.5

JCBD: Japanese diagnostic Criteria of Behçet's Disease; n: number of cases; NR: Not Reported

Objectives: In a well-defined Northern Spanish population-based cohort, the aim of this study was **a)** to estimate epidemiological variations, **b)** clinical domains and **c)** to compare our results with other regions.

Methods: Study of unselected all consecutive patients diagnosed with definitive or possible BD by expert rheumatologists between 1980 and 2020 in our health region. Two classification criteria were applied: **a)** International Study Group (ISG) for BD [1], and **b)** International Criteria for BD (ICBD) [2]. In addition, a literature review of Medline publications was carried out.

Results: In this study, from a total of 120 patients diagnosed with BD by expert opinion (58 women/62 men), 96 met ICBD and 59 ISG criteria. Mean age of the cohort at diagnosis was 37.6±13.8 years similar to other countries, as well as sex distribution.

Prevalence was higher than in most European populations regardless the diagnostic criteria applied: expert opinion (20.6), ICBD (16.5) or ISG (10.1) (TABLE 1). Incidence was lower (expert opinion: 0.022, ICBD: 0.018, ISG: 0.011). Clinical domains' frequency was in line with other regions except vascular and gastrointestinal involvement, which were lower.

Conclusion: BD's prevalence in Northern Spain is higher than in most European populations. These differences likely reflect a combination of true geographic variation, methodological artifacts as well as the easy access to Public Health System and its efficiency. In contrast, clinical phenotypes are similar to other regions.

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[1] *Lancet*. 1990; 335:1078-80

[2] *J Eur Acad Dermatol Venereol*. 2014; 28:338-47

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URINARY ALBUMIN-TO-CREATININE RATIO INDICATES NECROTIZING AND CRESCENTIC GLOMERULONEPHRITIS IN ANCA-ASSOCIATED VASCULITIS

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Background: Renal involvement is a common and severe complication of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) as it can cause acute kidney injury (AKI), end-stage renal disease (ESRD) or death.

Objectives: We have previously reported that elevated urinary albumin-to-creatinine ratio (uACR) correlates with rapid deterioration of kidney function in ANCA GN. Therefore, we here aimed to describe the association between proteinuric findings and histopathological diagnosis of necrotizing and crescentic ANCA GN in 50 urinary samples at admission and corresponding renal biopsies of patients with AAV.

Methods: A total number of 50 urinary samples at admission and corresponding renal biopsies with confirmed renal involvement of AAV were retrospectively included between 2015 till 2020 in a single-center observational study.

Results: Renal involvement of AAV revealed variable proteinuria ranging from low-range to nephrotic syndromes, however most patients presented with subnephrotic proteinuria. Severe deterioration of kidney function requiring RRT within 30 days after admission was associated with elevated levels of nonselective proteinuria, mostly attributed to albuminuria (uACR). Because we have previously shown that histologically confirmed ANCA GN with glomerular crescents and necrosis is associated with AKI and requirement of RRT during short-term disease course and elevated uACR levels were equally associated with AKI and requirement of RRT during the short-term course after disease onset, we next analyzed the association between uACR measurements at admission and histopathological findings within renal biopsies performed thereafter. Severely increased uACR levels >300mg/g correlated with reduction of normal glomeruli, attributed to increased glomerular crescents and necrosis. By contrast, no such association was observed for global sclerotic glomeruli, revealing that uACR reflects crescentic ANCA GN rather than adaptive glomerular hyperfiltration in chronic sclerosing stage. Since uACR levels could reflect both, either a specific renal involvement with necrotizing and crescentic ANCA GN or severity of systemic AAV disease, we next correlated uACR levels assessed at admission with extrarenal disease manifestation. We observed no association between uACR levels and extrarenal manifestation of AAV disease including pulmonary hemorrhage, skin involvement and BVAS assessment, suggesting that uACR levels reflected specific renal involvement in AAV. These observations were further confirmed by survival analysis for cumulative incidence of RRT during the short-term course of disease.

Conclusion: Early identification of patients who mostly benefit from aggressive immunosuppressive therapy is of clinical importance. Our observation that uACR levels at disease onset predict necrotizing and crescentic ANCA GN requires further investigation for therapeutic decision especially in patients with severe deterioration of kidney function.

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