

The epidemiology of Behçet's syndrome in England; a retrospective case control study nested within the Clinical Practice Research Datalink (CPRD) and CPRD linked Hospital Episodes Statistics (HES)

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Key Message:

This study identifies the prevalence / incidence of Behçet's in England and addresses risk factors associated with Behçet's and time to diagnosis from their onset. By recognizing these associated clinical phenotypes, there may be increased understanding of the disease and reduced diagnostic delay.

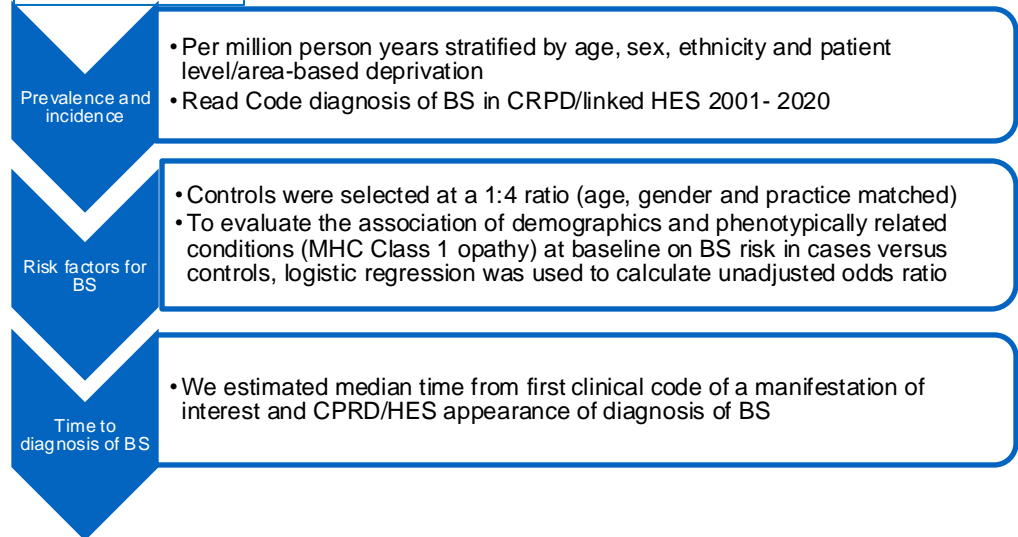
Introduction:

CPRD and linked HES provide a national resource to further explore the epidemiology of Behçet's syndrome (BS) in England. Conditions with both phenotypic and genetic overlap and demographic factors were examined to study the evolution of BS phenotype towards a confirmed diagnosis.

Objectives:

1. Annual incidence rate and prevalence per million person years were calculated stratified by demographics.
2. Risk factors for BS: demographics/ phenotypically linked conditions (MHC class 1 conditions).
3. Examine time to diagnosis of BS from first recorded symptom: mucocutaneous and systemic.

Methods:

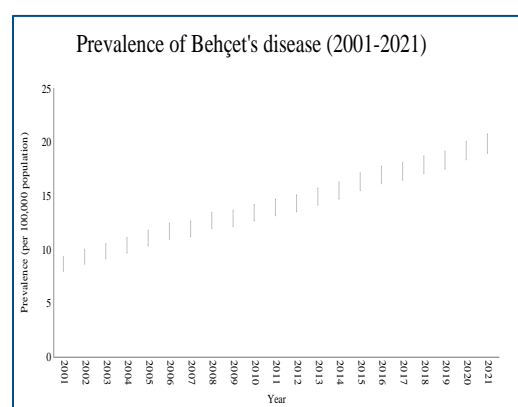
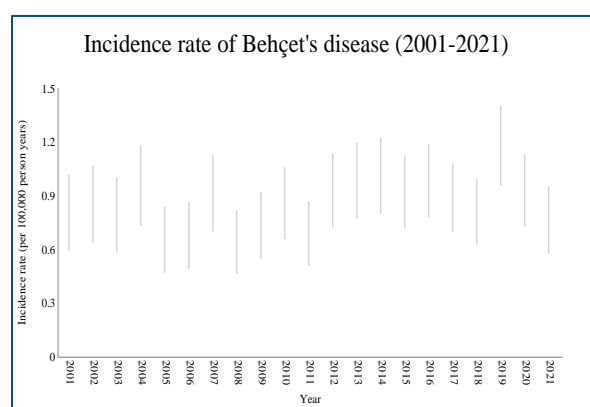


Results:

- Total of 4,810 cases of BS within the CRPD/linked HES database
- Incidence of BS stable; 0.78 (0.59 - 1.01)/100,000 PY in 2001 and 0.75 (0.58 - 0.95) in 2021. Prevalence rising; 8.62 (7.96 - 9.32) in 2001 to 19.80 (18.94 - 20.74) in 2021
- BS case matched (age/gender/practice registration date) to 10 controls: GU, uveitis and OU have the highest OR for BS, but seronegative spondyloarthritis, epididymitis, aneurysms, folliculitis and Crohn's were also associated with BS
- Median no. of days (IQR) for diagnosis of BS was shortest for GU 240 (63-745) and longest for enteropathic arthropathy 2494 (1268-3719)

Condition (Behçet N = 4234)	Number of Outcomes	Median (IQR) days
Aneurysms	21	814 (112.2-1595)
Ankylosing spondylitis	28	1111 (269-2754)
Cerebral venous thrombosis	4	834 (634-974)
Crohn's	47	662 (171.5-1328)
Enteropathic arthropathy	3	2494 (1268-3719)
Epididymitis	95	952 (363.5-2805.2)
Folliculitis	157	1547 (407-2862)
Genital ulcers	138	240 (63-745)
Oral ulcers	562	558 (141-1626)
Periodic fever	7	1400 (395-2480)
Psoriatic arthritis	20	741 (366-2167)
Reactive arthropathy	20	599 (316-2676)
Sweet syndrome	15	1262 (236.2-2599)
Uveitis	331	377.5 (107.5-1113)

Risk factor	Unadjusted Odds Ratio (95% CI)	P Value	Adjusted Odds Ratio (95% CI)	Adjusted P Value
Reactive arthropathy	17.28 (95% CI 5.77-63.07)	<0.001	16.78 (95% CI 5.59-61.32)	<0.001
Psoriatic arthritis	4.32 (95% CI 1.89-9.40)	<0.001	3.64 (95% CI 1.59-7.97)	0.001
Oral ulcers	24.73 (95% CI 20.16-30.51)	<0.001	24.73 (95% CI 20.14-30.54)	<0.001
Genital ulcers	142.13 (95% CI 71.56-336.20)	<0.001	155.08 (95% CI 77.90-367.40)	<0.001
Uveitis	44.14 (95% CI 31.25-64.16)	<0.001	43.00 (95% CI 30.39-62.59)	<0.001
Epididymitis	5.74 (95% CI 4.01-8.19)	<0.001	5.69 (95% CI 3.93-8.20)	<0.001
Aneurysms	4.32 (95% CI 1.89-9.40)	<0.001	3.28 (95% CI 1.42-7.25)	0.004
Folliculitis	2.44 (95% CI 1.91-3.10)	<0.001	2.46 (95% CI 1.92-3.12)	<0.001
Crohn's	6.01 (95% CI 3.69-9.74)	<0.001	5.69 (95% CI 3.48-9.25)	<0.001



Conclusions:

- Prevalence of BS appears higher than previously thought in the UK consistent with our previous study
- Misclassification with phenotypically and genetically related conditions may be a factor
- Risk of BS seems to be increased in those with a genetically linked condition
- Evident diagnostic delay can be avoided by increased recognition of key clinical manifestations
- Exploring such factors could be further facilitated by establishing a UK BS registry