INTRODUCTION

Review question / Objective: What are the causes of variability in clinical phenotype clustering in Behçet’s syndrome across different countries and cohorts?

Condition being studied: Behçet’s syndrome is a multisystem vasculitis and the presence of distinct clinical phenotypes with clustering of certain organ manifestations together is well-recognized. Differences in demographic features, treatment response, and possibly in inflammatory pathways involved in the pathogenesis of different phenotypes have been proposed. However, studies from different cohorts have shown variability in the phenotypes that were defined. We aimed to explore the causes of variability in clinical phenotype clustering across different countries and cohorts.

Information sources: The studies were searched by using Behçet, cluster, and factor analysis terms from chosen PubMed, EMBASE, and Cochrane Library.

INPLASY registration number: This protocol was registered with the International Platform of Registered Systematic Review and Meta-Analysis Protocols (INPLASY) on 27 February 2022 and was last updated on 27 February 2022 (registration number INPLASY202220119).
treatment response, and possibly inflammatory pathways involved in the pathogenesis of different phenotypes have been proposed. However, studies from different cohorts have shown variability in the phenotypes that were defined. We aimed to explore the causes of variability in clinical phenotype clustering across different countries and cohorts.

METHODS

Search strategy: Our search included all published studies in PubMed, EMBASE, and Cochrane Library from inception to before 12.02.2022. We also included only articles written in the English language.

Participant or population: Inclusion: patients with Behçet's syndrome (as diagnosed using any recognised diagnostic criteria) exclusion: editorials, comments, abstracts, or reviews without adequate raw data, articles written in languages other than English.

Intervention: Not applicable.

Comparator: Not applicable.

Study designs to be included: We will include all types of studies including Behçet patients.

Eligibility criteria: All studies reporting clusters in Behçet's syndrome will be included.

Information sources: The studies were searched by using Behçet, cluster, and factor analysis terms from chosen PubMed, EMBASE, and Cochrane Library.

Main outcome(s): The main outcome of this systematic review study will be to explain the reasons for different clusters in Behçet's syndrome.

Quality assessment / Risk of bias analysis: Two authors will independently assess the risk of bias using the Newcastle-Ottawa Quality Assessment tool. Any disagreements relevant to the risk of bias assessment will be resolved through discussion with a third author.

Strategy of data synthesis: A narrative or descriptive synthesis approach will be used.

Subgroup analysis: Subgroup analyses will be conducted according to the following four baseline characteristics: 1. Sex: Female, Male 2. Duration of disease 3. Patient population: Adult vs Pediatric 4. Statistical analysis method: hierarchical cluster analysis vs factor analysis

Sensitivity analysis: We will not plan to do a sensitivity analysis.

Language: Only articles written in English will be considered for inclusion.

Country(ies) involved: Turkey.

Keywords: Behçet, factor analysis, cluster.

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