Headache in Sickle Cell Disease: A Scoping Review Protocol

Registration: This protocol was formulated in-line with the PRISMA-ScR checklist (Preferred Reporting Items for Systematic reviews and Meta-Analysis extension for Scoping Reviews) and PRISMA-P (Preferred Reporting Items for Systematic Reviews and Meta-analysis Protocols). The protocol has been reviewed and will be submitted to Northwestern's Prism: https://prism.northwestern.edu/.

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Introduction

Rationale:

Headache is a common and often debilitating symptom of sickle cell disease (SCD). A significantly larger number of pediatric and adult patients with SCD experience severe and frequent headaches compared to healthy populations[1, 2]. The burden of this pain can manifest in a patient's life as negative impacts on their physical function, social and community participation, and emotional aspects of daily living[3]. In children, specifically, severe headache can result in absence from school, which may be especially impactful in their daily and future lives[4]. Although there are some studies that characterize headache prevalence and characteristics among SCD patients, they employ disparate definitions and methods. For example, one paper defines frequent headache as occurring at least once per week[1], and another defines the same term as headache occurring at least once per month[4]. There are very few studies that report the more specific characteristics of the headaches such as provoking factors, quality, region, severity, timing of episodes, and associated symptoms. Given the negative impact that headaches can have on the lives of SCD patients, it is important to gain a better understanding of the characteristics of headaches that occur in patients with SCD, identify gaps in knowledge, and use the information to explore potential new questions for future research.

Objectives:

This scoping review aims to compile evidence from existing literature about the prevalence, frequency, and characteristics of headache in sickle cell disease to strengthen current knowledge and to identify gaps in knowledge that may lead to new directions in research on headache in SCD.

Methods

Eligibility Criteria:

We will include studies with all numbers of human study subjects, including case reports with very few subjects, large case series, clinical trials, and others. Our selections will be limited to studies that contain data on patients with sickle cell disease, not sickle cell trait. Reports with data on both pediatric and adult patients will be included in our review. There will be no limitation with regards to publication date, however we will restrict our search to articles that are in the English language or that have been translated to English.

Information Sources:

MEDLINE (PubMed)

Cochrane Library (Wiley)

Embase (Elsevier)

Scopus (Elsevier)

Search Strategy:

The review team will work in collaboration with Annie Wescott, a research librarian from the Northwestern University Galter Health Sciences Library, to develop a search strategy to ensure comprehensive review of materials related to our topic. We will collaboratively select controlled vocabulary and keywords associated with the frequency and characteristics of headache in SCD and apply database-specific language and syntax for the following databases: MEDLINE (PubMed), Embase (Elsevier), and Scopus (Elsevier). The resulting list of abstracts will be deduplicated using a citation management software (EndNote) by Annie Wescott and then uploaded to the online abstract reviewing software Rayyan for blind review.

MEDLINE (PubMed) Search:

("Anemia, Sickle Cell"[Mesh] OR Sickle-cell-disease*[tiab] OR Sickle-cell-disorder*[tiab] OR Sickle-cell-anemia*[tiab] OR Sickle-cell-anaemia*[tiab] OR sickle-disease*[tiab] OR sickle-anemia*[tiab] OR sickle-anaemia*[tiab] OR Haemoglobin-S-Disease*[tiab] OR Haemoglobin-SS[tiab] OR Hemoglobin-SS[tiab] OR HbS-disease*[tiab] OR Hemoglobin-S-Disease*[tiab] OR SS-Disease*[tiab] OR drepanocytemia*[tiab] OR drepanocytic-anemia*[tiab] OR drepanocytic-anaemia*[tiab] OR drepanocytosis[tiab]) AND ("Headache"[Mesh] OR Headache* OR head-ache*[tiab] OR Head-pain*[tiab] OR Cranial-pain*[tiab] OR Cephalgia*[tiab] OR Cephalalgia*[tiab] OR Cephalodynia*[tiab] OR PQRST[tiab] OR symptom*[ti])

Study Selection:

The review team will be divided into three teams of two reviewers each. Prior to the formal grading of abstracts for selection in the scoping review, all reviewers will undergo a study orientation led by Aditi Joshi, who will introduce the purpose and methods of the study. During the orientation, all reviewers will become familiar with what a scoping review is designed to accomplish and the technology to be used for abstract review. After the orientation, all reviewers will grade 100 abstracts as a pilot round to test their collective understanding of the study methodology and techniques. During the formal review, each small team will separately evaluate equal portions of the total number of article abstracts that result from the searches. This will be done through Rayyan after our research librarian collaborator, Annie Wescott, completes the separate uploads of the abstracts onto the software for each team. If there are conflicting opinions within small teams about whether to include certain abstracts in the final review, a central review team will be consulted to resolve the conflicts. The central team will consist of reviewers Aditi Joshi, Dr. Philip Gorelick, and Dr. Yvonne Curran.

During formal review we will prioritize selecting studies that contain evidence about the "PQRST" of headaches in patients with SCD. We define the "PQRST" pain assessment method for headaches as follows:

P (provocation): aggravating and relieving factors

Q (quality): dull, throbbing, achy, sharp, etc.

R (region): unilateral, bilateral, frontal, etc.

S (severity): pain scale of 1 to 10, mild, moderate, severe

T (timing): frequency, number of headaches per week/ month, constant, intermittent, etc.

Study Evaluation:

Our goal is to use the "PQRST" headache characteristics to select studies that will allow us to create a comprehensive characterization of headaches in patients with SCD.

Data Management:

We will compile quantitative and qualitative data relating to the "PQRST" headache characteristics in SCD in a Microsoft Excel database to allow us to develop graphs and tables of the results in our manuscript. We will consider study limitations such as small sample sizes, missing characteristics of headache, disparate methods of data collection (e.g., prospective vs. retrospective methods) and definitions and grading of characteristics of headache, and generalizability of findings.

References

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