

Standardization of Coding Definitions for Sickle Cell Disease Complications: A Systematic Literature Review

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Abstract

Purpose: Sickle cell disease (SCD) affects all organ systems and is characterized by numerous acute and chronic complications and comorbidities. Standardized codes are needed for complications/comorbidities used in real-world evidence (RWE) studies that rely on administrative and medical coding. This systematic literature review was conducted to produce a comprehensive list of complications/comorbidities associated with SCD, along with their diagnosis codes used in RWE studies. **Methods:** A search in MEDLINE and Embase identified studies published from 2016-2023. Studies were included if they were conducted in US SCD populations and reported complications/comorbidities and respective International Classification of Diseases, Clinical Modification (ICD-CM) codes. All identified complications/comorbidities and codes were reviewed by a certified medical coding expert and hematologist. **Results:** Of 1,851 identified studies, 40 were included. The most reported complications/comorbidities were stroke, acute chest syndrome, pulmonary embolism, venous thromboembolism, vaso-occlusive crisis, and priapism. Most of the studies used ICD-9-CM codes (n=21), while some studies used ICD-10-CM codes (n=3) or both (n=16), depending on the study period. Most codes reported in literature were heterogeneous across complications/comorbidities. The medical coding expert recommended modifications for several conditions. **Conclusion:** While many studies we identified did not report their codes and were excluded from this review, the studies with codes exhibited diverse coding definitions. By providing a standardized set of diagnosis codes that were reported by studies and reviewed by a coding expert and hematologist, our review can serve as a foundation for accurately identifying complications/comorbidities in future research, and may reduce heterogeneity, enhance transparency, and improve reproducibility.

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