

STANDARD OF CARE FOR SICKLE CELL DISEASE IN CHILDREN AND ADOLESCENTS: A SCOPING REVIEW PROTOCOL

PADRÃO DE CUIDADO DA ANEMIA FALCIFORME PARA CRIANÇAS E ADOLESCENTES: PROTOCOLO DE REVISÃO DE ESCOPO

ESTÁNDAR DE ATENCIÓN PARA LA ANEMIA FALCIFORME EN NIÑOS Y ADOLESCENTES: PROTOCOLO DE REVISIÓN DE ALCANCE

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ABSTRACT

Introduction: Sickle cell anemia is a severe hereditary hemoglobinopathy, characterized by structural changes in red blood cells that result in episodes of vaso-occlusion, pain, and chronic tissue lesions, with significant repercussions on quality of life since childhood. In Brazil, it has a higher prevalence in the Northeast and Southeast regions, associated with African ancestry. Despite the advances resulting from neonatal screening and the National Policy for Comprehensive Care for People with Sickle Cell Disease, challenges persist related to access to specialized services, the training of health professionals, and the absence of standardized clinical protocols, which compromises the comprehensiveness of care, especially in primary care and emergency services. **Objective:** To map, in the global scientific literature, the standard of care for sickle cell anemia in children and adolescents, including clinical guidelines and care protocols. **Method:** This is a scoping review protocol conducted according to the methodology of the Joanna Briggs Institute and reported according to the recommendations of PRISMA-ScR. The sources of information will be PubMed/MEDLINE, Embase, Scopus, Web of Science, VHL, CINAHL, Cochrane Library, JBI Evidence Synthesis, and gray literature, such as Google Scholar, ResearchGate, and BDTD. The selection of studies will be carried out by two independent reviewers, with the support of the Rayyan software. **Expected results:** It is expected to identify and systematize guidelines, protocols, and recommendations related to the care of sickle cell anemia in children and adolescents, as well as to map knowledge gaps. **Conclusion:** The review may support the qualification of care practice and the strengthening of public policies aimed at the comprehensive care of this population.

Keywords: Anemia, Sickle Cell; Standard of Care; Comprehensive Health Care; Study Guides as Topic; Practice Guidelines as Topic.

RESUMO

Introdução: A anemia falciforme é uma hemoglobinopatia hereditária grave, caracterizada por alterações estruturais das hemácias que resultam em episódios de vaso-oclusão, dor e lesões teciduais crônicas, com repercussões significativas na qualidade de vida desde a infância. No Brasil, apresenta maior prevalência nas regiões Nordeste e Sudeste, associada à ancestralidade africana. Apesar dos avanços decorrentes da triagem neonatal e da Política Nacional de Atenção Integral às Pessoas com Doença Falciforme, persistem desafios relacionados ao acesso a serviços especializados, à capacitação dos profissionais de saúde e à ausência de protocolos clínicos padronizados, o que compromete a integralidade do cuidado, especialmente na atenção primária e nos serviços de urgência. **Objetivo:** Mapear, na literatura científica global, o padrão de cuidado da anemia falciforme em crianças e adolescentes, incluindo diretrizes clínicas e protocolos assistenciais. **Método:** Trata-se de um protocolo de revisão de escopo conduzido de acordo com a metodologia do Instituto Joanna Briggs e reportado conforme as recomendações do PRISMA-ScR. As fontes de informação serão PubMed/MEDLINE, Embase, Scopus, Web of Science, BVS, CINAHL, Cochrane Library, JBI Evidence Synthesis e literatura cinzenta, como Google Scholar, ResearchGate e BDTD. A seleção dos estudos será realizada por dois revisores independentes, com o apoio do software Rayyan. **Resultados esperados:** Espera-se identificar e sistematizar diretrizes, protocolos e recomendações relacionadas ao cuidado da anemia falciforme em crianças e adolescentes, bem como mapear lacunas do conhecimento. **Conclusão:** A revisão poderá subsidiar a qualificação da prática assistencial e o fortalecimento de políticas públicas voltadas ao cuidado integral dessa população.

Palavras-chave: Anemia Falciforme; Padrão de Cuidado; Assistência Integral à Saúde da Criança e do Adolescente; Guias como Assunto; Guias de Prática Clínica como Assunto.

RESUMEN

Introducción: La anemia falciforme es una hemoglobinopatía hereditaria grave, caracterizada por cambios estructurales en los glóbulos rojos que provocan episodios de vasooclusión, dolor y lesiones tisulares crónicas, con repercusiones significativas en la calidad de vida desde la infancia. En Brasil, tiene una mayor prevalencia en las regiones noreste y sureste, asociada a la ascendencia africana. A pesar de los avances derivados del cribado neonatal y la Política Nacional para la Atención Integral de las Personas con Anemia Falciforme, persisten desafíos relacionados con el acceso a servicios especializados, la formación de profesionales sanitarios y la ausencia de protocolos clínicos estandarizados, lo que compromete la atención integral, especialmente en atención primaria y servicios de urgencias. **Objetivo:** Mapear, en la literatura científica global, el estándar de atención para la anemia falciforme en niños y adolescentes, incluyendo guías clínicas y protocolos de cuidado. **Método:** Este es un protocolo de revisión de alcance realizado según la metodología del Instituto Joanna Briggs y reportado según las recomendaciones de PRISMA-ScR. Las fuentes de información serán PubMed/MEDLINE, Embase, Scopus, Web of Science, VHL, CINAHL, Cochrane Library, JBI Evidence Synthesis y literatura gris, como Google Scholar, ResearchGate y BDTD/NDLTD. La selección de estudios será realizada por dos revisores independientes, con el apoyo del software Rayyan. **Resultados esperados:** Se espera identificar y sistematizar directrices, protocolos y recomendaciones relacionados con el cuidado de la anemia falciforme en niños y adolescentes, así como mapear las lagunas de conocimiento. **Conclusión:** La revisión puede apoyar la cualificación de la práctica de atención y el fortalecimiento de políticas públicas orientadas a la atención integral de esta población.

Palabras-chave: Anemia de Células Falciformes; Nivel de Atención; Atención Integral de Salud; Guías como Asunto; Guías de Práctica Clínica como Asunto.

INTRODUCTION

Sickle cell anemia (SCA) is the homozygous (HbSS) and most severe form of sickle hemoglobinopathies, resulting from a homozygous mutation in the beta-globin gene (HBB, HbSS genotype). As a result of this structural alteration, red blood cells acquire a sickle shape, become rigid, and lose their ability to circulate freely, leading to vaso-occlusive phenomena, hemolysis, and chronic tissue injury. These manifestations compromise multiple organs and impact quality of life from early childhood, requiring constant and specialized care ^(1,2).

SCA represents one of the most prevalent genetic diseases worldwide, constituting a major global public health issue. Data from the Global Burden of Disease Study 2021 estimated 515,000 births of babies with sickle cell disease worldwide, of which 394,000 (76.5%) corresponded to the SS and S β^0 genotypes (severe form, including SCA), and 7.74 million people living with the disease globally, representing a 41.4% increase compared to 2000 ⁽³⁾.

The greatest burden is concentrated in Sub-Saharan Africa, accounting for 79% of births in 2021, but the disease is also relevant in the Americas and the Caribbean, with a 5.6% increase in birth incidence in Latin America and the Caribbean ⁽³⁾. In Brazil, it is estimated that between 25,000 and 50,000 people live with the homozygous or compound heterozygous form of the disease, with a higher prevalence in the Northeast and Southeast regions ^(4,5). This

distribution is strongly associated with African ancestry, reflecting historical and structural inequalities, as evidenced by data from the Ministry of Health ⁽⁶⁾.

Although there have been advances in the normative field, such as the implementation of neonatal screening in 2001 fundamental for promoting early diagnosis and reducing morbidity and mortality in the country and the creation of the National Policy for Comprehensive Care of People with Sickle Cell Disease (PNAIPDF), the challenges in implementing public policies and organizing care are still significant. Difficulties in accessing specialized services, a lack of clinical knowledge on the part of healthcare teams, underreporting of cases, the absence of standardized protocols, and the fragmentation of the healthcare network remain frequent barriers ^(1,5,7).

Children and adolescents with SCA require continuous and comprehensive monitoring, with attention directed beyond clinical manifestations, focusing on the psychosocial impacts of the disease. The Family Health Strategy (ESF) acts as the main gateway to the Unified Health System (SUS), being a strategic point for strengthening Primary Health Care (PHC) and playing a central role in the longitudinal follow-up of children and adolescents with SCA. However, many healthcare professionals still report feeling unprepared to deal with the condition, which compromises continuous and effective care ⁽¹⁾. Furthermore, emergency services lack

specific protocols for the management of sickle cell crises in childhood, resulting in inadequate or delayed approaches ⁽⁸⁾.

Furthermore, there is a scarcity of standardized clinical protocols aimed at the management of acute complications and systematized multidisciplinary care in emergency and primary care services, such as the management of sickle cell crises in childhood, resulting in inadequate or delayed approaches ^(7,8). This scenario highlights the need for intersectoral actions that promote equity in the care of this population, especially in light of the racial and social inequalities that permeate the history of the disease in Brazil ⁽⁴⁾.

A preliminary literature search was conducted to verify the existence of previously published scoping reviews or systematic reviews on the topic. This exploratory search was conducted in databases such as PubMed/MEDLINE and the Virtual Health Library (VHL), using terms related to sickle cell disease, healthcare, clinical guidelines, and care protocols.

Review studies addressing specific aspects of sickle cell disease, such as clinical management, complications, quality of life, and health policies, were identified. However, no scoping reviews were found focusing on mapping clinical guidelines, care protocols, and standards of care specifically directed at children and adolescents with sickle cell disease, highlighting a knowledge gap and reinforcing the relevance of the present protocol.

The present review is justified by the convergence of gaps that compromise the quality and continuity of care, considering: the fragmentation of care across primary, emergency, and specialized services; the scarcity of standardized protocols for the management of SCA in childhood; and the high mortality risk in children with sickle cell anemia. This highlights the need to understand the coverage of existing protocols and to provide a basis for the integration of care actions directed at this population.

Given this context, this scoping review protocol aims to map the standard of care for sickle cell anemia in children and adolescents in the global scientific literature, including clinical guidelines and care protocols, in order to identify advances, gaps, and possibilities for improving healthcare.

METHODS

Study type, protocol, and registration

This is a scoping review protocol conducted based on the methodology proposed by the Joanna Briggs Institute (JBI) for the conduct of scoping reviews. The development of the protocol will follow the recommendations of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR) ⁽⁹⁻¹⁰⁾. The protocol was previously registered on the Open Science Framework (OSF) under the access DOI: <https://doi.org/10.17605/OSF.IO/HB6VP>.

The choice of a scoping review is justified as it is a methodological approach designed to map and synthesize the key concepts

and available evidence in a given field of knowledge, as well as to examine the extent of research and identify gaps in the scientific literature⁽¹¹⁾.

Research question

The formulation of the research question and the structuring of the inclusion criteria were based on the PCC mnemonic (Population, Concept, and Context), defined as: (1) Population (P) – Children and adolescents (aged 0 to under 18 years) with a confirmed diagnosis of sickle cell anemia (HbSS genotype); (2) Concept (C) – Standard of care, encompassing clinical practice guidelines, care protocols, technical manuals, and institutional recommendations; (3) Context (C) – Global scientific literature and official documents, with no restrictions regarding geographical location or level of healthcare.

Based on these elements, the guiding research question was defined as: “What exists in the global literature regarding the standard of care for sickle cell anemia, including clinical guidelines and protocols, for children and adolescents?”.

Eligibility criteria

The eligibility criteria were defined based on the PCC framework and document typology. Publications focusing on the population of children and adolescents aged 0 to under 18 years diagnosed with SCA, encompassing the HbSS genotype, will be included. Regarding the concept, studies and documents addressing the standard of care for SCA will be considered, encompassing clinical practice guidelines, care

protocols, technical manuals, public policies, and health recommendations. Concerning the context, the search will cover the global scientific literature, without any geographical delimitation or restriction regarding the level of healthcare. Primary studies with quantitative, qualitative, and mixed-methods approaches, secondary studies, and institutional grey literature produced by government agencies or scientific societies will be eligible, with no filters applied for publication period or language.

Conversely, documents that address sickle cell disease or other hemoglobinopathies broadly and generically, without presenting a specific sub-analysis or stratification for SCA, will be excluded. Publications in which care protocols or guidelines are not clearly and sufficiently described for data extraction will also be disregarded, as well as editorials, letters to the editor, conference abstracts, commentaries, book chapters, and articles based exclusively on expert opinion without normative or institutional endorsement. Serão incluídos na análise final apenas estudos com texto integral disponível.

If any eligible document is not directly accessible in full text within the databases or institutional repositories, up to three attempts will be made to contact the corresponding author via email over a three-week period. If there is no response or the file is not made available after this period, the study will be definitively excluded due to full-text unavailability⁽¹¹⁾.

Information sources

Searches will be conducted in the following indexed bibliographic databases: PubMed/MEDLINE, Excerpta Medica Database (EMBASE), Scopus, Web of Science, Virtual Health Library (VHL), Latin American and Caribbean Health Sciences Literature (LILACS via VHL), Cumulative Index to Nursing and Allied Health Literature (CINAHL), Cochrane Library, and JBI Evidence Synthesis. Additionally, grey literature and institutional repositories will be consulted to map theses, dissertations, and guidelines not formally published, encompassing Google Scholar (evaluating the first 200 results sorted by relevance), ResearchGate, and the Brazilian Digital Library of Theses and Dissertations (BDTD)/Networked Digital Library of Theses and Dissertations (NDLTD).

To expand the retrieval of relevant articles, backward and forward citation searches will be conducted by examining the reference lists of the included studies and identifying

Table 1 – Preliminary search strategy in PubMed

IS*	Search strategy	Retrieved articles
PubMed	(Child OR "Child, Preschool" OR "Infant" OR "Newborn" OR "ADOLEC" OR Adolescent) AND ("Anemia, Sickle Cell") AND ("Standard of Care" OR "Evidence-Based Practice" OR "Guidelines as Topic" OR "Practice Guideline" OR "eHealth Strategies" OR "Clinical Protocols" OR "Health Planning Guidelines" OR "Practice Guidelines as Topic" OR "Guideline Adherence")	259

*IS: Information source.

Source: Prepared by the authors.

(2) Development and application of the final search strategies combining controlled descriptors and keywords using the Boolean operators AND and OR, with appropriate qualifier adaptations for each database. The

subsequent citations. Institutional repositories and academic networks will also be consulted to locate full texts. No filters for publication date or study type will be applied, ensuring a comprehensive approach.

Search strategy

For the search strategy, a three-step approach will be employed: (1) an initial exploratory search in PubMed, VHL, and EMBASE, accessed via the CAPES Periodicals Portal through the Federated Academic Community (CAFe), aimed at identifying controlled descriptors in the Medical Subject Headings (MeSH), Health Sciences Descriptors (DeCS), and Emtree vocabularies, as well as keywords and free-text terms present in the titles and abstracts of related publications. The structured preliminary strategy (Table 1) for PubMed combines the following blocks of terms:

detailed syntax adapted for each of these sources is described in Appendix I; and (3) a complementary search in the grey literature, including Google Scholar, ResearchGate, the Brazilian Digital Library of Theses and

Dissertations (BDTD), and the Networked Digital Library of Theses and Dissertations (NDLTD).

Study selection

The records retrieved from all information sources will be exported to a reference manager and imported into the Rayyan Intelligent Systematic Review software, where the automated and manual identification and removal of duplicates will be performed ⁽¹²⁾. The selection of publications will occur in two blind and independent phases (blind mode), conducted by two reviewers. The first phase will consist of screening and reading the titles and abstracts of all retrieved records.

The second phase will comprise the full-text reading of all publications pre-selected in the previous stage to confirm final eligibility based on the previously established criteria (full-text screening). To ensure standardization and consistency in the application of the inclusion and exclusion criteria between the reviewers, a prior calibration step will be conducted on a random sample of 25 retrieved publications.

Prior to the start of formal screening, the two reviewers will conduct a calibration test on a random sample of 25 publications. A minimum absolute agreement rate of 75% between the reviewers will be established as a satisfactory threshold to validate the calibration. If the achieved percentage is lower, the eligibility criteria will be discussed, aligned, and re-evaluated on a new sample of 25 records until the established target is reached ⁽¹¹⁾.

Any disagreements between the two reviewers arising during Phase 1 or Phase 2 will initially be addressed through consensus meetings for a joint re-evaluation of the document. If the divergence persists after discussion, the definitive inclusion or exclusion of the study will be deliberated and decided by a third reviewer.

Data extraction and analysis

The extraction of information from the eligible publications will be conducted independently by the two reviewers using a standardized form structured according to the JBI guidelines (Appendix II). The data collection instrument will undergo a prior pilot test on two selected studies to verify its clarity, comprehensiveness, and applicability in capturing the variables of interest.

Data synthesis and presentation of results

Data analysis will be conducted descriptively, with thematic categorization of the extracted contents, allowing the identification of recurrences, gaps, and contextual variations among the mapped evidence.

EXPECTED RESULTS

The results will be presented through tables, charts, flowcharts, and thematic maps, according to the nature of the identified evidence. These visual representations will be accompanied by a narrative synthesis aimed at contextualizing the findings, highlighting the most relevant evidence, and identifying knowledge gaps. The discussion of the results will consider their implications for policymaking, the improvement of care

practices, and the management of health services, especially in the care of children and adolescents with SCA.

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Conflict of interest statement

None to declare.

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All content underlying the article is contained within the manuscript.

Authorship criteria (author contributions)

Conception and design of the study; or acquisition, analysis, or interpretation of research

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Drafting or critical revision of the manuscript for important intellectual content: Luiz, LDS; Santos, FP; Fonseca, BS; Rissi, GP; Higarashi, IH.

Final approval of the version to be published: Luiz, LDS; Santos, FP; Fonseca, BS; Caparroz, HF; Rissi, GP; Higarashi, IH.

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APPENDICES

Appendix I: Data extraction instrument

Objective	To map, in the global literature, the standard of care for sickle cell anemia, including clinical guidelines and protocols, for children and adolescents.		
	P	C	C
Extraction	Children and adolescents	Standard of care for sickle cell anemia	Global level of healthcare
Conversion	Child Child, Preschool Infant Newborn ADOLEC Adolescent	Standard of Care Evidence-Based Practice Guidelines as Topic Practice Guideline Clinical Protocols Health Planning Guidelines Practice Guidelines as Topic Guideline Adherence Anemia, Sickle Cell	-
Combination	MESH Child Children Child, Preschool Preschool Child Children, Preschool Preschool Children Infant Infant* Infant, Newborn Infant*, Newborn Newborn Infant* Neonate* Newborn* Adolescent Adolescent* Adolescence Adolescent*, Female Female Adolescent* Adolescent*, Male Male Adolescent* Youth* Teen* Teenager*	MESH Standard of Care Care Standard* Standard* of Care Evidence-Based Practice Evidence Based Practice Evidence-Based Health Care Evidence-Based Health Care* Health Care*, Evidence-Based Evidence Based Healthcare Evidence-Based Healthcare* Healthcare, Evidence-Based Healthcare*, Evidence-Based Evidence Based Management, Healthcare Evidence Based Management, Health Care Evidence-Based Health Care Management Evidence-Based Healthcare Management Guidelines as Topic Guidelines as Topics Practice Guideline Clinical Practice Guideline Clinical Guideline* Treatment Guideline Intervention Guideline Therapeutic Guideline Guideline Clinical Protocols Clinical Protocol Protocol*, Clinical Treatment Protocol* Protocol*, Treatment Clinical Research Protocol Protocol*, Clinical Research Research Protocol, Clinical Clinical Research Protocol* Research Protocol*, Clinical Health Planning Guidelines Guideline*, Health Planning Health Planning Guideline Planning Guideline*, Health Guideline* for Health Planning Recommendation*, Health Planning Health Planning Recommendation Health Planning Recommendation* Planning Recommendation*, Health Recommendation, Health Planning	MESH -

		Practice Guidelines as Topic Clinical Guidelines as Topic Best Practice* Guideline Adherence Adherence, Guideline Institutional Adherence Adherence, Institutional Policy Compliance Compliance, Policy Protocol Compliance Compliance, Protocol Anemia, Sickle Cell Anemias, Sickle Cell Sickle Cell Anemia* Hemoglobin S Disease Disease, Hemoglobin S Hemoglobin S Disease* Sickle Cell Disease Cell Disease*, Sickle Sickle Cell Disease* Sickle Cell Disorder* Cell Disorder*, Sickle Sickle Cell Disorder Sickling Disorder Due to Hemoglobin S HbS Disease Sickle Cell Anemia	
	EMTREE 'child' OR 'children' OR 'child' OR 'child, preschool' OR 'pre-school child' OR 'infant' OR 'newborn' OR 'teenager' OR 'adolescent'	EMTREE 'standard of care' OR 'standards of care' OR 'evidence based' OR 'evidence-based practice' OR 'evidence based practice' OR 'guidelines' OR 'guidelines as topic' OR 'practice guidelines' OR 'practice guidelines as topic' OR 'practice guideline' OR 'clinical protocols' OR 'clinical protocol' OR 'protocol' OR 'guideline adherence' 'anemia sickle cell in infancy and childhood' OR 'anemia sickle cell in infant and child' OR 'anaemia, sickle cell' OR 'anemia, sickle cell' OR 'sickle anaemia' OR 'sickle anemia' OR 'sickle cell anaemia' OR 'sickle cell disease' OR 'sickle cell disorder' OR 'SS disease (sickle cell)' OR 'sickle cell anemia'	EMTREE -
	DECS Criança OR Child OR Niño OR Enfant OR "Pré-Escolar" OR "Child, Preschool" OR Prescolar OR "Enfant d'âge préscolaire" OR Lactente OR Infant OR Lactante OR Nourrisson OR "Recém-Nascido" OR "Infant, Newborn" OR "Recién Nacido" OR "Nouveau-né" OR Adolescente OR Adolescent Adolescente OR	DECS "Padrão de Cuidado" OR "Standard of Care" OR "Nivel de Atención" OR "Norme de soins" OR "Prática Clínica Baseada em Evidências" OR "Evidence- Based Practice" OR "Práctica Clínica Basada en la Evidencia" OR "Pratique factuelle" OR "Guias como Assunto" OR "Guidelines as Topic" OR "Guías como Asunto" OR "Recommandations comme sujet" OR "Diretrizes para o Planejamento em Saúde" OR "Health Planning Guidelines" OR "Directrices para la Planificación en Salud" OR "Directives de santé publique" OR "Guia de Prática Clínica" OR "Practice Guideline" OR "Guía de Práctica Clínica" OR "Guide de bonnes	DECS

	Adolescent	pratiques” OR Guia OR Guideline OR Guia OR Directives OR “Protocolos Clínicos” OR “Clinical Protocols” OR “Protocolos Clínicos” OR “Protocoles cliniques” OR “Guias de Prática Clínica como Assunto” OR “Practice Guidelines as Topic” OR “Guías de Práctica Clínica como Asunto” OR “Guides de bonnes pratiques cliniques comme sujet” OR “Fidelidade a Diretrizes” OR “Guideline Adherence” OR “Adhesión a Directriz” OR “Adhésión aux directives” AND “Anemia Falciforme” OR “Anemia, Sickle Cell” OR “Anemia de Células Falciformes” OR Drépanocytose	
Construction	MESH (Child) OR (Children) OR (Child, Preschool) OR (Preschool Child) OR (Children, Preschool) OR (Preschool Children) OR (Infant) OR (Infant*) OR (Infant, Newborn) OR (Infant*, Newborn) OR (Newborn Infant*) OR (Neonate*) OR (Newborn*) OR (Adolescent) OR (Adolescent*) OR (Adolescence) OR (Adolescent*, Female) OR (Female Adolescent*) OR (Adolescent*, Male) OR (Male Adolescent*) OR (Youth*) OR (Teen*) OR (Teenager*)	MESH (Standard of Care) OR (Care Standard*) OR (Standard* Care) OR (Evidence-Based Practice) OR (Evidence Based Practice) OR (Evidence-Based Health Care) OR (Evidence Based Health Care) OR (Evidence-Based Health Care*) OR (Health Care*, Evidence-Based) OR (Evidence-Based Healthcare) OR (Evidence Based Healthcare) OR (Evidence-Based Healthcare*) OR (Healthcare, Evidence-Based) OR (Healthcare*, Evidence-Based) OR (Evidence Based Management, Healthcare) OR (Evidence Based Management, Health Care) OR (Evidence Based Health Care Management) OR (Evidence Based Healthcare Management) OR (Guidelines as Topic) OR (Guidelines as Topics) OR (Practice Guideline) OR (Clinical Practice Guideline) OR (Clinical Guideline*) OR (Treatment Guideline) OR (Intervention Guideline) OR (Therapeutic Guideline) OR (Guideline) OR (Clinical Protocols) OR (Clinical Protocol) OR (Protocol*, Clinical) OR (Treatment Protocol*) OR (Protocol*, Treatment) OR (Clinical Research Protocol) OR (Protocol*, Clinical Research) OR (Research Protocol, Clinical) OR (Clinical Research Protocol*) OR (Research Protocol*, Clinical) OR (Health Planning Guidelines) OR (Guideline*, Health Planning) OR (Health Planning Guideline) OR (Planning Guideline*, Health) OR (Guideline* Health Planning) OR (Recommendation*, Health Planning) OR (Health Planning Recommendation) OR (Health Planning Recommendation*) OR (Planning Recommendation*, Health) OR (Recommendation, Health Planning) OR (Practice Guidelines as Topic) OR	MESH

		(Clinical Guidelines as Topic) OR (Best Practice*) OR (Guideline Adherence) OR (Adherence, Guideline) OR (Institutional Adherence) OR (Adherence, Institutional) OR (Policy Compliance) OR (Compliance, Policy) OR (Protocol Compliance) OR (Compliance, Protocol) (Anemia, Sick Cell) OR (Anemias, Sick Cell) OR (Sickle Cell Anemias) OR (Hemoglobin S Disease) OR (Disease, Hemoglobin S) OR (Hemoglobin S Diseases) OR (Sickle Cell Disease) OR (Hemoglobin S Disease) OR (Disease, Hemoglobin S) OR (Hemoglobin S Diseases) OR (Sickle Cell Disease) OR (Cell Disease, Sick) OR (Cell Diseases, Sick) OR (Sickle Cell Diseases) OR (Sickle Cell Disorders) OR (Cell Disorder, Sick) OR (Cell Disorders, Sick) OR (Sickle Cell Disorder) OR (Sickling Disorder Due to Hemoglobin S) OR (HbS Disease) OR (Sickle Cell Anemia)	
	EMTREE 'child' OR 'children' OR 'child' OR 'child, preschool' OR 'pre-school child' OR 'infant' OR 'newborn' OR 'teenager' OR 'adolescent'	EMTREE 'standard of care' OR 'standards of care' OR 'evidence based' OR 'evidence-based practice' OR 'evidence based practice' OR 'guidelines' OR 'guidelines as topic' OR 'practice guidelines' OR 'practice guidelines as topic' OR 'practice guideline' OR 'clinical protocols' OR 'clinical protocol' OR 'protocol' OR 'guideline adherence' AND 'anemia sickle cell in infancy and childhood' OR 'anemia sickle cell in infant and child' OR 'anaemia, sickle cell' OR 'anemia, sickle cell' OR 'sickle anaemia' OR 'sickle anemia' OR 'sickle cell anaemia' OR 'sickle cell disease' OR 'sickle cell disorder' OR 'SS disease (sickle cell)' OR 'sickle cell anemia'	EMTREE
	DECS Criança OR Child OR Niño OR Enfant OR “Pré-Escolar” OR “Child, Preschool” OR Prescolar OR “Enfant d’âge pré-scolaire” OR Lactente OR Infant OR Lactante OR Nourrisson OR “Recém-Nascido” OR “Infant, Newborn” OR “Recién Nacido” OR “Nouveau-né” OR Adolescente OR Adolescent Adolescente OR Adolescent	DECS “Padrão de Cuidado” OR “Standard of Care” OR “Nivel de Atención” OR “Norme de soins” OR “Prática Clínica Baseada em Evidências” OR “Evidence-Based Practice” OR “Práctica Clínica Basada en la Evidencia” OR “Pratique factuelle” OR “Guias como Assunto” OR “Guidelines as Topic” OR “Guías como Asunto” OR “Recommandations comme sujet” OR “Diretrizes para o Planejamento em Saúde” OR “Health Planning Guidelines” OR “Directrices para la Planificación en Salud” OR “Directives de santé publique” OR “Guia de Prática Clínica” OR “Practice Guideline” OR “Guía de Práctica Clínica” OR “Guide de bonnes pratiques” OR Guia OR Guideline OR Guía OR Directives OR “Protocolos	DECS

	Clínicos” OR “Clinical Protocols” OR “Protocolos Clínicos” OR “Protocolos cliniques” OR “Guias de Prática Clínica como Assunto” OR “Practice Guidelines as Topic” OR “Guías de Práctica Clínica como Asunto” OR “Guides de bonnes pratiques cliniques comme sujet” OR “Fidelidade a Diretrizes” OR “Guideline Adherence” OR “Adhesión a Directriz” OR “Adhésion aux directives” AND “Anemia Falciforme” OR “Anemia, Sickle Cell” OR “Anemia de Células Falciformes” OR Drépanocytose	
Database	Search Strategy	Retrieved Articles
PUBMED	18/09/2025 ((((((((((((((((((((((Child[MeSH Terms])) OR (Children[Title/Abstract])) OR (Child, Preschool[MeSH Terms])) OR (Preschool Child[Title/Abstract])) OR (Children, Preschool[Title/Abstract])) OR (Preschool Children[Title/Abstract])) OR (Infant[MeSH Terms])) OR (Infant*[Title/Abstract])) OR (Infant, Newborn[MeSH Terms])) OR (Infant*, Newborn[Title/Abstract])) OR (Newborn Infant*[Title/Abstract])) OR (Neonate*[Title/Abstract])) OR (Newborn*[Title/Abstract])) OR (Adolescent[MeSH Terms])) OR (Adolescent*[Title/Abstract])) OR (Adolescence[Title/Abstract])) OR (Adolescent*, Female[Title/Abstract])) OR (Female Adolescent*[Title/Abstract])) OR (Adolescent*, Male[Title/Abstract])) OR (Male Adolescent*[Title/Abstract])) OR (Youth*[Title/Abstract])) OR (Teen*[Title/Abstract])) OR (Teenager*[Title/Abstract])) AND (((((((((((((((((((((((Standard of Care[MeSH Terms])) OR (Care Standard*[Title/Abstract])) OR (Standard* of Care[Title/Abstract])) OR (Evidence-Based Practice[MeSH Terms])) OR (Evidence Based Practice[Title/Abstract])) OR (Evidence Based Health Care[Title/Abstract])) OR (Evidence-Based Health Care*[Title/Abstract])) OR (Health Care*, Evidence-Based[Title/Abstract])) OR (Evidence Based Healthcare[Title/Abstract])) OR (Evidence-Based Healthcare*[Title/Abstract])) OR (Healthcare, Evidence-Based[Title/Abstract])) OR (Healthcare*, Evidence-Based[Title/Abstract])) OR (Evidence Based Management, Healthcare[Title/Abstract])) OR (Evidence Based Management, Health Care[Title/Abstract])) OR (Evidence Based Health Care Management[Title/Abstract])) OR (Evidence Based Healthcare Management[Title/Abstract])) OR (Guidelines as Topic[MeSH Terms])) OR (Guidelines as Topics[Title/Abstract])) OR (Practice Guideline[MeSH Terms])) OR (Clinical Practice Guideline[Title/Abstract])) OR (Clinical Guideline*[Title/Abstract])) OR (Treatment Guideline[Title/Abstract])) OR (Intervention Guideline[Title/Abstract])) OR (Therapeutic Guideline[Title/Abstract])) OR (Guideline[MeSH Terms])) OR (Clinical Protocols[MeSH Terms])) OR (Clinical Protocol[Title/Abstract])) OR (Protocol*, Clinical[Title/Abstract])) OR (Treatment Protocol*[Title/Abstract])) OR (Protocol*, Treatment[Title/Abstract])) OR (Clinical Research Protocol[Title/Abstract])) OR (Protocol*, Clinical Research[Title/Abstract])) OR (Research Protocol, Clinical[Title/Abstract])) OR (Clinical Research Protocol*[Title/Abstract])) OR (Research Protocol*, Clinical[Title/Abstract])) OR (Health Planning Guidelines[MeSH Terms])) OR (Guideline*, Health Planning[Title/Abstract])) OR	332

105

	TS=(Guideline Adherence)) OR TS=(Adherence, Guideline)) OR TS=(Institutional Adherence)) OR TS=(Adherence, Institutional)) OR TS=(Policy Compliance)) OR TS=(Compliance, Policy)) OR TS=(Protocol Compliance)) OR TS=(Compliance, Protocol) #1 (((((((((((((((((((((TS=(Child)) OR TS=(Children)) OR TS=(Child, Preschool)) OR TS=(Preschool Child)) OR TS=(Children, Preschool)) OR TS=(Preschool Children)) OR TS=(Infant)) OR TS=(Infant*)) OR TS=(Infant, Newborn)) OR TS=(Infant*, Newborn)) OR TS=(Newborn Infant*)) OR TS=(Neonate*)) OR TS=(Newborn*)) OR TS=(Adolescent)) OR TS=(Adolescent*)) OR TS=(Adolescence)) OR TS=(Adolescent*, Female)) OR TS=(Female Adolescent*)) OR TS=(Adolescent*, Male)) OR TS=(Male Adolescent*)) OR TS=(Youth*)) OR TS=(Teen*)) OR TS=(Teenager*))	
Cochrane	18/09/2025 #1 MeSH descriptor: [Child] explode all trees #2 MeSH descriptor: [Child, Preschool] explode all trees #3 MeSH descriptor: [Infant] explode all trees #4 MeSH descriptor: [Adolescent] explode all trees #5 MeSH descriptor: [Standard of Care] explode all trees #6 MeSH descriptor: [Evidence-Based Practice] explode all trees #7 MeSH descriptor: [Guidelines as Topic] explode all trees #8 MeSH descriptor: [Clinical Protocols] explode all trees #9 MeSH descriptor: [Practice Guidelines as Topic] explode all trees #10 MeSH descriptor: [Guideline Adherence] explode all trees #11 MeSH descriptor: [Anemia, Sick Cell] explode all trees #12 (#1 OR #2 OR #3 OR #4) AND (#5 OR #6 OR #7 OR #8 OR #9 OR #10) AND #11	1
CINAHL	18/09/2025 ((Child) OR (Child, Preschool) OR (Infant) OR (Newborn) OR (ADOLEC) OR (Adolescent)) AND ((Standard of Care) OR (Evidence-Based Practice) OR (Guidelines as Topic) OR (Practice Guideline) OR (eHealth Strategies) OR (Clinical Protocols) OR (Health Planning Guidelines) OR (Practice Guidelines as Topic) OR (Guideline Adherence)) AND (Anemia, Sick Cell)	488
EMBASE	18/09/2025 #19 AND #20 AND #21 #21 'anemia sickle cell'/exp OR 'sickle cell anemia'/exp OR 'sickle cell disease'/exp #20 'guideline'/exp OR 'practice guideline'/exp OR 'clinical protocol'/exp #19 'child'/exp OR 'children'/exp OR 'adolescent'/exp OR 'infant'/exp OR 'teenager'/exp	769
SCOPUS	18/09/2025 ('child' OR 'children' OR 'child' OR 'child, preschool' OR 'pre-school child' OR 'infant' OR 'newborn' OR 'teenager' OR 'adolescent') AND ('standard of care' OR 'standards of care' OR 'evidence based' OR 'evidence-based practice' OR 'evidence based practice' OR 'guidelines' OR 'guidelines as topic' OR 'practice guidelines' OR 'practice guidelines as topic' OR 'practice guideline' OR 'clinical protocols' OR 'clinical protocol' OR 'protocol' OR 'guideline adherence') AND ('anemia sickle cell in infancy and childhood' OR 'anemia sickle cell in infant and child' OR 'anaemia, sickle cell' OR 'anemia, sickle cell' OR 'sickle anaemia' OR 'sickle anemia' OR 'sickle cell anaemia' OR 'sickle cell disease' OR 'sickle cell disorder' OR 'sickle cell anemia')	20
BVS	18/09/2025 (criança OR child OR niño OR enfant OR "Pré-Escolar" OR "Child, Preschool" OR preescolar OR "Enfant d'âge préscolaire" OR lactente OR infant OR lactante OR nourrisson OR "Recém-Nascido" OR "Infant, Newborn" OR "Recién Nacido" OR "Nouveau-né" OR adolescente OR adolescent OR adolescente OR adolescent) AND ("Padrão de Cuidado" OR "Standard of Care" OR "Nível de Atención" OR "Norme de soins" OR "Prática Clínica Baseada em Evidências" OR	333

	"Evidence-Based Practice" OR "Práctica Clínica Basada en la Evidencia" OR "Pratique factuelle" OR "Guias como Assunto" OR "Guidelines as Topic" OR "Guías como Asunto" OR "Recommandations comme sujet" OR "Diretrizes para o Planejamento em Saúde" OR "Health Planning Guidelines" OR "Directrices para la Planificación en Salud" OR "Directives de santé publique" OR "Guia de Prática Clínica" OR "Practice Guideline" OR "Guía de Práctica Clínica" OR "Guide de bonnes pratiques" OR guia OR guideline OR OR "Protocolos Clínicos" OR "Clinical Protocols" OR "Protocolos clínicos" OR "Guías de Práctica Clínica como Assunto" OR "Practice Guidelines as Topic" OR "Guías de Práctica Clínica como Asunto" OR "Guides de bonnes pratiques cliniques comme sujet" OR "Fidelidade a Diretrizes" OR "Guideline Adherence" OR "Adhesión a Directriz" OR "Adhésion aux directives") AND ("Anemia Falciforme" OR "Anemia, Sickle Cell" OR "Anemia de Células Falciformes" OR drépanocytose)	
JB1	18/09/2025 "Child" OR "Child, Preschool" OR Infant OR "Infant, Newborn" OR Adolescent; "Standard of Care" OR "Evidence Based Practice" OR "Guidelines as Topic" OR "Practice Guideline" OR "Guideline" OR "Clinical Protocols" OR "Health Planning Guidelines" OR "Practice Guidelines as Topic" OR "Guideline Adherence"; "Anemia, Sickle Cell"	1
TOTAL		2049

Appendix II: Search strategy

AXIS	EXTRACTION VARIABLE
STUDY IDENTIFICATION	Title
	Authors/Issuing body
	Publication year
	Origin/Country
	Language
	Objectives
	Study/document type
	Methods
POPULATION	Profile of subjects and target of actions
CONCEPT	Preventive and prophylactic measures
	Management of acute events and emergencies
	Disease-modifying and maintenance therapies
	Multidisciplinary care and care pathway
CONTEXT	Global care setting and healthcare systems
SYNTHESIS	Evidence, impact, and gaps