

Clinical and laboratory risk factors for sickle cell retinopathy and maculopathy: a scoping review of the current evidence

Rajani P. Brandsen,^{1,2} Roselie M.H. Diederer,¹ Gizem Kocabas,¹ Erfan Nur,^{2,3} Arjan Malekzadeh,⁴ Reinier O. Schlingemann^{1,5} and Bart J. Biemond²

¹Department of Ophthalmology, Amsterdam UMC, University of Amsterdam, Amsterdam, the Netherlands; ²Department of Hematology, Amsterdam UMC, University of Amsterdam, Amsterdam, the Netherlands; ³Sanquin Research and Landsteiner Laboratory, Department of Blood Cell Research, Amsterdam, the Netherlands; ⁴Medical Library, Amsterdam UMC, University of Amsterdam, Amsterdam, the Netherlands and ⁵Department of Ophthalmology, University of Lausanne, Jules-Gonin Eye Hospital, Fondation Asile des Aveugles, Lausanne, Switzerland

Correspondence: R.P. Brandsen
r.p.brandsen@amsterdamumc.nl

Received: September 3, 2024.
Accepted: December 6, 2024.
Early view: December 19, 2024.

<https://doi.org/10.3324/haematol.2024.286420>

©2025 Ferrata Storti Foundation

Published under a CC BY-NC license



Appendix A – Search strategies

Database(s): Ovid MEDLINE(R) ALL 1946 to May 22, 2023	
#	Searches
1	exp Anemia, Sickle Cell/ or exp Sickle Cell Trait/
2	(sickle adj3 cell adj3 (an?emia* or disease* or trait* or patholog* or disorder* or syndrom*)).ti,ab,kf.
3	((Hb or Hemoglobin) adj3 ("S disease*" or "SS disease")).ti,ab,kf.
4	or/1-3
5	exp Retinal Degeneration/ or exp Retinal Diseases/
6	((retinopath* or retin* or macul* or ocular or eye) adj4 (dystroph* or degenerat* or disease* or path* or abnormal*)).ti,ab,kf.
7	(retinopath* or maculopath*).ti,ab,kf.
8	or/5-7
9	4 and 8
10	exp Risk Factors/ or exp Incidence/ or exp Mass Screening/ or exp "Severity of Illness Index"/
11	((("fetal h?emoglobin" or h?emoglobin or incidence or (ophthalmic or systemic or biomarker* or laboratory or chemical* or risk or health* or vulnerab* or susceptib* or disease* or disorder* or clinical* or environmental* or genet* or genom* or epidemiolog* or prognostic or predict*)) adj3 (factor* or correlate* or indicator* or profile*)).ti,ab,kf.
12	((disease or retinopath* or retin* or macul* or ocular or eye or ophthalmological or strategy) adj3 (assess* or screen* or examination)).ti,ab,kf.
13	or/10-12
14	9 and 13

Database(s): Embase Classic+Embase 1947 to 2023 May 22	
#	Searches
1	exp sickle cell anemia/ or exp sickle cell trait/
2	("sickle cell" adj3 (an?emia* or disease* or trait* or patholog* or disorder* or syndrom*)).ti,ab,kw.

3	((Hb or Hemoglobin) adj3 ("S disease*" or "SS disease")).ti,ab,kw.
4	or/1-3
5	exp retina degeneration/ or exp retina disease/
6	((retinopath* or retin* or macul* or ocular or eye) adj4 (dystroph* or degenerat* or disease* or path* or abnormal*)).ti,ab,kw.
7	(retinopath* or maculopath*).ti,ab,kw.
8	or/5-7
9	4 and 8
1 0	exp risk factor/ or exp incidence/ or exp screening/ or exp "severity of illness index"/
1 1	((("fetal h?emoglobin" or h?emoglobin or incidence or (ophthalmic or systemic or biomarker* or laboratory or chemical* or risk or health* or vulnerability or susceptib* or disease* or disorder* or clinical* or environmental* or genet* or genom* or epidemiolog* or prognostic or predict*)) adj3 (factor* or correlate* or indicator* or profile*)).ti,ab,kw.
1 2	((disease or retinopath* or retin* or macul* or ocular or eye or ophthalmological or strategy) adj3 (assess* or screen* or examination)).ti,ab,kf.
1 3	or/10-12
1 4	9 and 13

Scopus

TITLE-ABS-KEY-AUTH (("sickle cell") W/3 (an?emia* OR disease* OR trait* OR patholog* OR disorder* OR syndrom*) OR ((hb OR hemoglobin) W/3 ("S disease*" OR "SS disease"))) AND TITLE-ABS-KEY-AUTH (retinopath* OR maculopath* OR ((retin* OR macul* OR ocular OR eye) W/4 (dystroph* OR degenerat* OR disease* OR patholog* OR abnormal*))) AND TITLE-ABS-KEY-AUTH ((("fetal h?emoglobin" OR h?emoglobin OR incidence OR (ophthalmic OR systemic OR biomarker* OR laboratory OR chemical* OR risk OR health* OR vulnerability OR susceptib* OR disease* OR disorder* OR clinical* OR environmental* OR genet* OR genom* OR epidemiolog* OR prognostic OR predict*)) W/3 (factor* OR correlate* OR indicator* OR profile*)) OR ((disease OR retinopath* OR retin* OR macul* OR ocular OR eye OR ophthalmological OR strategy) W/3 (assess* OR screen* OR examination)))