

Appendix Table 1. Search Strategies

Database	Search String	Total Returns
MEDLINE	(“Anemia, Sickle Cell”[MeSH] OR “sickle cell”[tiab]) AND (“Hydroxyurea”[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT “meta-analysis”[pt]	5708
	(“Hydroxyurea”[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab]) AND (“Drug Toxicity”[MeSH] OR "drug toxicity"[tiab] OR toxicity[tiab] OR harm[tiab] OR "adverse event"[tiab] OR neoplasms[mh] OR neoplasm*[tiab] OR malignancy[tiab] OR cancer[tiab] OR "Leg Ulcer"[MeSH] OR "leg ulcer"[tiab] OR "Nausea"[MeSH] OR nausea[tiab] OR vomit*[tiab] OR "Alopecia"[MeSH] OR "hair loss"[tiab] OR myelosuppression[tiab] OR (delay[tiab] AND (development*[tiab] OR growth[tiab])) OR teratogen*[tiab] OR "Safety"[MeSH] OR safety[tiab] OR "Leukemia"[MeSH] OR leukemia[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT “meta-analysis”[pt]	
	("Anemia, Sickle Cell"[MeSH] OR "sickle cell"[tiab]) AND ("Hydroxyurea"[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab] OR "Phenylbutyrates"[MeSH] OR "sodium phenylbutyrate"[tiab] OR arginine[mh] OR butyrate[mh] OR "arginine butyrate"[tiab] OR "decitabine"[Substance Name] OR decitabine[tiab] OR "Azacitidine"[MeSH] OR azacitidine[tiab] OR azacytidine[tiab] OR "5-azacitidine"[tiab] OR "5-azacytidine"[tiab] OR "Penicillins"[MeSH] OR penicillin[tiab] OR "Folic Acid"[MeSH] OR "folic acid"[tiab] OR folate[tiab] OR "Vaccines"[MeSH] OR "Vaccination"[MeSH] OR vaccine*[tiab] OR "iron chelation"[tiab] OR ("Iron"[MeSH] OR iron[tiab] OR Fe[tiab]) AND ("Chelation Therapy"[MeSH] OR "Therapeutics"[MeSH] OR "chelation therapy"[tiab])) OR "Nutrition Therapy"[MeSH] OR "nutrition therapy"[tiab] OR "nutrition counseling"[tiab] OR ("Pain"[MeSH] AND management[tiab]) OR "pain management"[tiab] OR "Dental Care for Chronically Ill"[MeSH] OR "chronic transfusion"[tiab] OR (chronic[tiab] AND transfusion[tiab]) OR "Bone Marrow Transplantation"[MeSH] OR "bone marrow transplant*" [tiab] OR treatment[tiab] OR "Patient Care Management"[MeSH] OR "case management"[tiab]) AND ("Health Policy"[MeSH] OR "Ethics"[MeSH] OR ethic*[tiab] OR "Delivery of Health Care"[MeSH] OR "delivery of health care"[tiab] OR "health care delivery"[tiab] OR "Social Support"[MeSH] OR "Psychology"[MeSH] OR psychology[tiab] OR bias[tiab] OR "Psychology"[MeSH] OR "Costs and Cost Analysis"[MeSH] OR cost[tiab] OR "Health behavior"[MeSH] OR communication[tiab] OR Barrier*[tiab] OR "patient satisfaction"[tiab] OR "Comorbidity"[MeSH] OR comorbidity[tiab] OR "Depression"[MeSH] OR depression[tiab] OR "socioeconomic status"[tiab] OR "Social Support"[MeSH Major Topic] OR "family support"[tiab] OR "Education"[MeSH] OR education[tiab] OR "Insurance, Health, Reimbursement"[MeSH] OR "Quality of Health Care"[MeSH] OR "quality of care"[tiab] OR "practice pattern"[tiab] OR "disease severity"[tiab] OR burden[tiab] OR "cognitive ability"[tiab] OR respect[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT “meta-analysis”[pt]	
	(“Anemia, Sickle Cell”[MeSH] OR “sickle cell”[tiab] OR "Thrombocythemia, Hemorrhagic"[MeSH] OR “essential thrombocythemia”[tiab]) AND (“Hydroxyurea”[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT “meta-analysis”[pt]	

	("Anemia, Sickle Cell"[MeSH] OR "sickle cell"[tiab]) AND ("Health Policy"[MeSH] OR "Ethics"[MeSH] OR ethic*[tiab] OR "Delivery of Health Care"[MeSH] OR "delivery of health care"[tiab] OR "health care delivery"[tiab] OR "Social Support"[MeSH] OR "Psychology"[MeSH] OR psychology[tiab] OR bias[tiab] OR "Psychology"[MeSH] OR "Costs and Cost Analysis"[MeSH] OR cost[tiab] OR "Health behavior"[MeSH] OR communication[tiab] OR Barrier*[tiab] OR "patient satisfaction"[tiab] OR "Depression"[MeSH] OR depression[tiab] OR "socioeconomic status"[tiab] OR "Social Support"[MeSH Major Topic] OR "family support"[tiab] OR "Education"[MeSH] OR education[tiab] OR "Insurance, Health, Reimbursement"[MeSH] OR "Quality of Health Care"[MeSH] OR "quality of care"[tiab] OR "practice pattern"[tiab] OR burden[tiab] OR respect[tiab] OR religion[MeSH] OR spirituality[tiab] OR religion[tiab] OR "internal-external control"[MeSH]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT "meta-analysis"[pt]	
EMBASE	("Anemia, Sickle Cell"[MeSH] OR "sickle cell"[tiab]) AND ("Hydroxyurea"[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT "meta-analysis"[pt]	5009
	("Hydroxyurea"[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab]) AND ("Drug Toxicity"[MeSH] OR "drug toxicity"[tiab] OR toxicity[tiab] OR harm[tiab] OR "adverse event"[tiab] OR neoplasms[mh] OR neoplasm*[tiab] OR malignancy[tiab] OR cancer[tiab] OR "Leg Ulcer"[MeSH] OR "leg ulcer"[tiab] OR "Nausea"[MeSH] OR nausea[tiab] OR vomit*[tiab] OR "Alopecia"[MeSH] OR "hair loss"[tiab] OR myelosuppression[tiab] OR (delay[tiab] AND (development*[tiab] OR growth[tiab])) OR teratogen*[tiab] OR "Safety"[MeSH] OR safety[tiab] OR "Leukemia"[MeSH] OR leukemia[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT "meta-analysis"[pt]	
	("Anemia, Sickle Cell"[MeSH] OR "sickle cell"[tiab]) AND ("Hydroxyurea"[MeSH] OR hydroxyurea[tiab] OR hydra[tiab] OR hydroxycarbamide[tiab] OR "Phenylbutyrates"[MeSH] OR "sodium phenylbutyrate"[tiab] OR arginine[mh] OR butyrate[mh] OR "arginine butyrate"[tiab] OR "decitabine"[Substance Name] OR decitabine[tiab] OR "Azacitidine"[MeSH] OR azacitidine[tiab] OR azacytidine[tiab] OR "5-azacitidine"[tiab] OR "5-azacytidine"[tiab] OR "Penicillins"[MeSH] OR penicillin[tiab] OR "Folic Acid"[MeSH] OR "folic acid"[tiab] OR folate[tiab] OR "Vaccines"[MeSH] OR "Vaccination"[MeSH] OR vaccine*[tiab] OR "iron chelation"[tiab] OR ("Iron"[MeSH] OR iron[tiab] OR Fe[tiab]) AND ("Chelation Therapy"[MeSH] OR "Therapeutics"[MeSH] OR "chelation therapy"[tiab]) OR "Nutrition Therapy"[MeSH] OR "nutrition therapy"[tiab] OR "nutrition counseling"[tiab] OR ("Pain"[MeSH] AND management[tiab]) OR "pain management"[tiab] OR "Dental Care for Chronically Ill"[MeSH] OR "chronic transfusion"[tiab] OR (chronic[tiab] AND transfusion[tiab]) OR "Bone Marrow Transplantation"[MeSH] OR "bone marrow transplant*"[tiab] OR treatment[tiab] OR "Patient Care Management"[MeSH] OR "case management"[tiab]) AND ("Health Policy"[MeSH] OR "Ethics"[MeSH] OR ethic*[tiab] OR "Delivery of Health Care"[MeSH] OR "delivery of health care"[tiab] OR "health care delivery"[tiab] OR "Social Support"[MeSH] OR "Psychology"[MeSH] OR psychology[tiab] OR bias[tiab] OR "Psychology"[MeSH] OR "Costs and Cost Analysis"[MeSH] OR cost[tiab] OR "Health behavior"[MeSH] OR communication[tiab] OR Barrier*[tiab] OR "patient satisfaction"[tiab] OR "Comorbidity"[MeSH] OR comorbidity[tiab] OR "Depression"[MeSH] OR depression[tiab] OR "socioeconomic status"[tiab] OR "Social Support"[MeSH Major Topic] OR "family support"[tiab] OR "Education"[MeSH] OR education[tiab] OR "Insurance, Health, Reimbursement"[MeSH] OR "Quality of Health Care"[MeSH] OR "quality of care"[tiab] OR "practice pattern"[tiab] OR "disease severity"[tiab] OR burden[tiab] OR "cognitive ability"[tiab] OR respect[tiab]) AND English[lang] NOT (animal[mh] NOT human[mh]) NOT review[pt] NOT "meta-analysis"[pt]	

	(((('sickle cell anemia'/exp) OR ('sickle cell':ti,ab) OR ('thrombocytopenia'/exp) OR ('essential thrombocytopenia':ti,ab)) AND (('hydroxyurea'/exp) OR (hydroxyurea:ti,ab) OR (hydrea:ti,ab) OR (hydroxycarbamide:ti,ab))) AND english:la) NOT ((review:it) OR ('meta analysis':it)) AND [humans]/lim	
	(((('sickle cell anemia'/exp) OR ('sickle cell':ti,ab)) AND (('health policy'/exp) OR ('ethics'/exp) OR ('delivery of healthcare':ti,ab) OR ('health care delivery':ti,ab) OR ('social support'/exp) OR ('social support':ti,ab) OR ('psychology'/exp) OR (psychology:ti,ab) OR ('health care cost'/exp) OR (cost:ti,ab) OR ('health behavior'/exp) OR ('communication':ti,ab) OR (barrier:ti,ab) OR ('patient satisfaction'/exp) OR ('depression'/exp) OR (depression:ti,ab) OR ('social class'/exp) OR ('socioeconomic status':ti,ab) OR ('education'/exp) OR (education:ti,ab) OR ('health insurance'/exp) OR ('health insurance':ti,ab) OR ('health care quality'/exp) OR (burden:ti,ab) OR (respect:ti,ab) OR ('religion'/exp) OR (religion:ti,ab) OR ('control'/exp))) AND english:la) NOT ((review:it) OR ('meta analysis':it)) AND [humans]/lim	
TOXLine	((hydroxyurea OR hydrea OR hydroxycarbamide OR 127-07-1 [rn]) NOT ("in vitro" OR (animals) OR pregnant OR (pregnancy) OR reproductive OR (child) OR pediatric OR (adolescent))) AND (eng [la]) AND (BIOSIS [org] OR CIS [org] OR CRISP [org] OR EPIDEM [org] OR FEDRIP [org] OR IPA [org] OR MTGABS [org] OR PubMed [org] OR RISKLINE [org] OR TSCATS [org])	1602
CINAHL	(MM hydroxyurea or TX hydroxyurea or TX hydrea or TX hydroxycarbamide) and (MM ("drug toxicity" or "adverse drug event" or neoplasms or "leg ulcer" or nausea or alopecia) or TX ("drug toxicity" or "adverse drug event" or neoplasms or "leg ulcer" or nausea or alopecia or malignancy or cancer or vomiting or "hair loss" or myelosuppression))	
	(MM "anemia, sickle cell" or TX "sickle cell" or TX thrombocytopenia) and (MM (Hydroxyurea or arginine or vaccines or "dental care for chronically ill" or "case management") or TX (folate or "iron chelation" or "chronic transfusion"))	
	((MM "anemia, sickle cell" or TX "sickle cell" or TX thrombocytopenia)) and (MM ("practice pattern" or "insurance, health, reimbursement" or education or "support, psychosocial" or depression or communication or "health behavior" or "cost and cost analysis" or "health care costs" or "health care delivery" or ethics or "health policy" thrombocytopenia) or TX ("social support" or bias or barriers or "socioeconomic status" or support or "disease severity"))	
Handsearching	–	5