

**Table 1. Common Complications and Treatment Options in Sickle Cell Disease**

COMPLICATION (PATHOPHYSIOLOGY)	SIGNS AND SYMPTOMS	DIFFERENTIAL DIAGNOSES AND DIAGNOSTIC TESTS	TREATMENT OPTIONS
Dactylitis or hand-foot syndrome (Vaso-occlusion)	Symmetric swelling of hands and/or feet Age 6 mos to 5 yrs	Cellulitis, osteomyelitis CBC, x-ray	Fluids, pain management
Infection (Splenic dysfunction, immunocompromised)	Fever, chills, cough, dysuria, flank pain, headache	Sepsis, UTI, pneumonia, meningitis, osteomyelitis CBC with diff, retic, blood cx, UA, LP, CXR	Empiric abx Immunization PCN prophylaxis until age 5 yrs
Pain episodes or "crisis" (Vaso-occlusion)	Pain to extremities, back, chest, abdomen, etc. Typical "crisis pain"	Infection, osteomyelitis, dehydration, costochondritis, AVN, trauma, gallstones, etc. CBC with diff, retic, CMP, UA	Hydration Analgesia
Acute chest syndrome (infection, infarction, embolism)	Chest pain, dyspnea, cough, fever, wheezing, hypoxia	Pneumonia, trauma, costochondritis CBC with diff, retic, blood cx, CXR, ABG	ABX, transfusion, IS, oxygen, ventilator support, inhaled NO
Stroke or CVA (Vaso-occlusion, vasculopathy)	Paralysis, aphasia, hemiparesis, monoparesis, seizure, headache	Meningitis, viral syndrome, migraine CBC with diff, retic, type and cross match, CT, MRI/MRA of brain, LP	Simple or exchange transfusion, monitor for cerebral edema, analgesia, rehabilitation
Splenic sequestration (Trapping of sickled RBCs)	Abdominal swelling and pain, pallor, jaundice, fatigue, dyspnea, tachycardia, hypotension	Gallstones, hepatitis, pyelonephritis, hyperhemolysis, aplastic anemia CBC with diff, retic, CMP, abd ultrasound, abd CT	Transfusion, fluid resuscitation, consider splenectomy
Aplastic anemia or "aplastic crisis" (Suppression of RBC progenitors)	Fatigue, pallor, tachycardia, dyspnea	Aplastic anemia, DBA, TEC, folate or vit B12 def, splenic sequestration CBC with diff, retic, parvovirus titers, epo level, ferritin, Vit B12 and folate levels	Observation, transfusion, erythropoietin, folate supplements
Priapism (Vaso-occlusion)	Prolonged, painful erection	CBC with diff, retic, UA, urine cx, type and screen	Hydration, analgesia, warm baths, corporal aspiration/irrigation, transfusion, shunting procedure, ephedrine, androgen block
Gallstones (Bilirubin from increased hemolysis)	RUQ abd pain, post prandial nausea, worsening jaundice	Pancreatitis, gastritis, splenic sequestration, pain episode CBC with diff, retic, CMP, abd ultrasound or CT	Analgesia, Cholecystectomy if symptomatic.
Leg ulcer (Poor peripheral circulation)	Difficult to heal leg ulcer, swelling, pain	Cellulitis, infection, trauma CBC with diff, retic, x-ray, bone scan	Wet to dry dressing, leg elevation, PT, consider abx and debridement
Nephropathy (Vaso-occlusion)	Enuresis, proteinuria, hypertension, renal insufficiency or renal failure	CBC with diff, retic, CMP, UA, 24 hrs urine for creatinine clearance and microalbuminuria, renal ultrasound	ACE inhibitors, other antihypertensives

**Key:** UTI—urinary tract infection, CBC with diff—complete blood cell count with white blood cell differential, Retic—reticulocyte count, Cx—cultures, UA—urinalysis, Mos—months, Yrs—years, CXR—chest x-ray, Abx—antibiotics, PCN—penicillin, AVN—avascular necrosis, CMP—complete metabolic panel, Abd—abdomen, IS—incentive spirometry, ABG—arterial blood gas, NO—nitric oxide, CVA—cerebrovascular accident, CT—computed tomography, MRI—magnetic resonance imaging, MRA—magnetic resonance angiogram, LP—lumbar puncture, RBC—red blood cell, DBA—diamond blackfan anemia, TEC—transient erythroblastopenia of childhood, Epo—erythropoietin, RUQ—right upper quadrant, PT—physical therapy, ACE—angiotensin-converting enzyme.

**Note:** Diagnostic studies and treatment options should be based on clinical findings. Not all listed options need to be applied in every circumstance.