TABLE 1. Spectrum of acute sickle cell hepatopathy in childhood

Classification	Clinical features	Serum AST/ALT	Serum total bilirubin	Management
Vasoocclusive hepatic crisis	RUQ pain, nausea, fever, hepatomegaly	< 300 U/L	≤ 15 mg/dL, mainly conjugated	Intravenous hydration, analgesics
Hepatic sequestration crisis	RUQ pain, hepatomegaly, anemia (acute decrease > 2 g/dL), thrombocytopenia	< 300 U/L	≤ 24 mg/dL, mainly conjugated	Supportive care with (exchange) transfusion therapy
Sickle cell intrahepatic cholestasis	RUQ pain, nausea fever, rapidly progressing to liver failure with coagulopathy and renal impairment	> 1000 U/L	Strikingly elevated, mainly conjugated	Intensive care, exchange transfusion, correction of coagulopathy
Benign extreme hyperbilirubinemia	Minimal or no symptoms	< 500 U/L	20 - 58 mg/dL, half conjugated	Spontaneous resolution in 2-8 weeks

[1]Adapted from Shah *et al.*, Banerjee *et al.,* and Buchanan & Glader. Abbreviations: ALT, alanine transaminase; AST, aspartate transaminase; RUQ, right upper quadrant.