Hemin and sickle cell disease-associated acute chest syndrome development

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Acute chest syndrome (ACS) is a complication of sickle cell disease that is characterized by sudden pain and difficulty breathing. Sickle cell disease can also cause red blood cells to suddenly breakdown and release their contents, which may trigger the onset of ACS.

In this issue of the *Journal of Clinical Investigation*, Solomon Fiifi Ofori-Acquah and colleagues at Emory University asked if hemin, a product released by red blood cells during lysis, triggers ACS in a mouse model of sickle cell disease.

They found that hemin injection caused labored breathing, acute lung injury and rapid death in sickle mice, but not in control mice.

They determined that hemin acts through the immune receptor TLR4, since blocking or deleting this receptor prevented symptoms of hemin injection in sickle mice.

Hemopexin, a protein that sequesters hemin was an effective treatment against hemin-induced lung injury both before and after symptom onset.

These data suggest that therapies aimed at targeting hemin and/or blocking TLR4 signaling may be valuable approaches for treatment or prevention of acute chest syndrome.

More information: Extracellular hemin crisis triggers acute chest syndrome in sickle mice, *J Clin Invest*. DOI: 10.1172/JCI64578