

Lipoprotein Abnormalities Contribute to Sickle Cell Disease Pathology

PRESS RELEASE **UPDATED: JUN 23, 2017**

Washington, DC, June 23, 2017 (Newswire.com) - An article published in *Experimental Biology and Medicine* (Volume 242, Issue 12, June, 2017) links imbalances in lipoprotein metabolism with vaso-occlusive events in patients with sickle cell disease (SCD). The study, led by Dr. Eric Soupene in Dr. Frans Kuypers' laboratory at the Children's Hospital Oakland Research Institute (CHORI) in Oakland CA, identified high density lipoprotein (HDL) metabolites in SCD plasma that promote inflammation and may reduce the effectiveness of current therapies.

The red blood cells in patients with SCD contain a mutated hemoglobin, the protein that carries oxygen throughout the body. SCD patients also have abnormalities in cholesterol metabolites (plasma cholesterol and lipoproteins) and other plasma components such as acute phase reactants, inflammatory mediators, cell free hemoglobin, and heme. In SCD, the red blood cells (RBCs) become inflexible and stick to the walls of blood vessels, which prevents the delivery of oxygen to cells and tissues. These vaso-occlusive events are further exacerbated by abnormalities in plasma components. The standard treatment for vaso-occlusive events is replacement of the sickle RBCs with normal RBCs. However, RBC exchange therapy does not normalize alterations in plasma components. Thus, the

📄 PRESS CONTACT

📄 ABOUT EXPERIMENTAL BIOLOGY AND MEDICINE

Experimental Biology and Medicine is a journal dedicated to the publication of multidisciplinary and interdisciplinary research in the biomedical sciences. The journal was first established in 1903.

majority of the plasma present after RBC exchange therapy will form a proinflammatory environment that can reduce the effectiveness of the treatment.

Previous studies have demonstrated that the proinflammatory environment of the blood in patients with SCD reduces the activity of high density lipoprotein (HDL), the so called 'good' cholesterol particle. In the current study, Dr. Soupene and colleagues characterized HDL metabolites in SCD plasma and their effect on endothelial cell function. SCD plasma exhibited alterations in the size distribution, but not the total level, of HDL particles. SCD plasma was depleted of the pre- β particle, which is essential for reverse cholesterol transport. This observation may partially explain the altered plasma cholesterol content observed in SCD patients. HDL isolated from SCD plasma upregulated PTX3, a protein that is predictive for the length of inflammatory episodes in SCD, in endothelial cells. The addition of the heme-scavenger hemopexin (Hx) blocked PTX3 upregulation by SCD plasma. Collectively, these findings link lipoprotein alterations to SCD pathology, and suggest that replacement of plasma and RBCs, whole blood transfusion, may be a more effective treatment for vaso-occlusive events. According to Dr. Soupene, "RBC exchange therapy, a well-accepted transfusion treatment of SCD patients, may have to be re-evaluated. Replacing only the red blood cells underestimates the importance alterations in the plasma compartment."

Dr. Steven R. Goodman, Editor-in-Chief of *Experimental Biology and Medicine*, said, "Soupene and colleagues have provided an important new view on treatment of vaso-occlusive episodes. They clearly demonstrate an altered HDL particle size distribution in SCD plasma that leads to increased inflammatory response. Their work points to consideration of whole blood transfusion, as an alternative to RBC exchange therapy, as a treatment for vaso-occlusive episodes."

About Experimental Biology and Medicine

Experimental Biology and Medicine is a journal dedicated to the publication of multidisciplinary and interdisciplinary research in the biomedical sciences. The journal was first established in 1903. *Experimental Biology and Medicine* is the journal of the Society of Experimental Biology and Medicine. To learn about the benefits of society membership, visit www.sebm.org. If you are interested in publishing in the journal, please visit <http://ebm.sagepub.com>.

Disclaimer: *Newswire is not responsible for the accuracy of news releases posted to Newswire by contributing institutions or for the use of any information through the Newswire platform.*

Source: Experimental Biology and Medicine

 Share on Facebook

 Share on Twitter

 

 **Categories:**

Healthcare and Medical News,
Research, Medical Research

 **Tags:**

ebm, experimental biology and
medicine, lipoprotein, sickle cell,
sickle cell disease, sickle cell
disease pathology