

Prevention of sickle cell disease progression in adult mice

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Sickle cell disease (SCD) is characterized by abnormal hemoglobin, which alters the shape of red blood cells, resulting in poor blood flow and eventual death. In developed countries, the survival rate in children with SCD is high due to early medical intervention. However, the risk of adverse effects dramatically increases in SCD patients during early adulthood, and the factors that underlie adult progression of the disease are poorly understood.

A new study in *JCI Insight* reports the results of a longitudinal study of SCD model mice that links impaired activity of the antioxidant regulator Nrf2 to intravascular red blood cell destruction and other adverse SCD-associated effects. Solomon Ofori-Acquah and colleagues at the University of Pittsburgh found that the severity of hemolytic anemia, vascular inflammation, and lung injury increases with age in SCD mice. Pharmacological activation of Nrf2 in young animals improved survival and lessened age-related adverse effects. Additionally, expression of Nrf2 in non-blood cells was crucial for protection against tissue damage.

Together, these results suggest that Nrf2 augmentation should be further explored for treating SCD.

More information: Samit Ghosh et al. Nonhematopoietic Nrf2 dominantly impedes adult progression of sickle cell anemia in mice, *JCI Insight* (2016). [DOI: 10.1172/jci.insight.81090](https://doi.org/10.1172/jci.insight.81090)

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