

## **Vascular Effects of Hemolysis**

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Chronic hemolytic red blood cell disorders as diverse as sickle cell disease, thalassemia, spherocytosis, and some other red blood cell enzyme hemoglobin disorders have been linked to occasional high blood pressure in the lungs, leg ulceration, blood clots, strokes and painful involuntary erection of the penis. The causes of these problems are complex and incompletely understood, but several clues are available. Red blood cells contain mainly hemoglobin, the red pigment that makes red blood cells red and carries oxygen from the lungs to our organs and tissues. The breakdown of old or abnormal red blood cells can release the contents of the red cells into plasma, the liquid part of blood surrounding the blood cells. Several of these substances can affect the functions of the blood vessel wall, especially hemoglobin which acts as a strong oxidant when it escapes the red blood cell. Free hemoglobin depletes nitric oxide (NO), a gas normally produced by the blood vessel wall that helps to keep the vessels wide open. Chronically low levels of nitric oxide over many years can lead to high blood pressure in the lungs, which can become a serious problem in some people. Several natural mechanisms in the bloodstream help to protect against free hemoglobin, and its breakdown products, including heme and free iron. Other red blood cell products, including the ATP, ADP and the enzyme arginase, also appear to contribute to the risk of blood vessel disorders and blood clots. It seems clear that adults with sickle cell anemia should have echocardiography at least once in adulthood to screen for high blood pressure in the lungs. It should be a consideration in other people with severe chronic hemolytic anemia, especially if they have high serum levels of lactate dehydrogenase (LDH), or progressive shortness of breath with exertion or exercise. Current evidence suggests that the risk of these complications is increased by all the factors that raise the risk of heart attack and stroke in the general population. People with chronic hemolytic anemias, like the rest of us, should practice the healthy lifestyles that appear to reduce risk of heart attacks and strokes: healthy diets, exercise, avoid smoking, and carefully manage high blood pressure, high cholesterol and diabetes.