Anemia is the condition in which there is a decrease in red blood cells and volume. As a result, oxygen-rich cells carry less and less of that essential element through the bloodstream. People are fatigued and tired and their organs don’t function optimally. It would stand to reason that an excess of red cells in the circulating blood, called polycythemia, might actually be favorable to health and well being, but alas, it is not. Polycythemia carries a series of problems and complications that make it a serious disorder to underwrite. 

There are two main types of polycythemia, primary and secondary. In primary, also known as polycythemia vera, bone marrow stem cells undergo slow and steady reproduction that gradually results in a taking-over of the bone marrow. Not only are red cells overproduced, but the infection-fighting white blood cells as well as platelets (which are clotting helpers) are over-created as well. Polycythemia vera is usually genetic, and 95 percent of people affected have a mutation in the signaling molecule JAK2. About 60 percent of those affected are men, and the usual age of onset is between 50 and 60 years of age. 

While “normal” levels of hemoglobin and other blood products may vary within a range, that spread elevates significantly in polycythemia. Normal hemoglobin and hematocrit levels in the 13 to 15 and 40 to 46 range respectively go to 16 and 50 and often beyond. White blood cells—generally fewer than 10,000 in the absence of infection—routinely go over that. Platelets, with an upper normal limit of 400,000, go to 600,000 and over. It starts to get pretty crowded in the bone marrow and blood stream, and serious complications develop.

While there are no symptoms early on, those affected begin to manifest problems related to an expanded blood volume and increased blood viscosity, or thickness. Headache, dizziness, ringing in the ears, blurred vision and fatigue develop. Itching is a common sign related to the increased number of one type of the white blood cells overproduced (basophils) secreting excess histamine. Blood clogs the smaller blood vessels of the nose, and epistaxis (nosebleed) is not an uncommon finding.

When red cells more or less overrun the body, the spleen begins to store them up, and an enlarged spleen is a hallmark of diagnosis. The spleen may burst, causing a surgical emergency. Thrombosis ( clotting) is the most common complication and is the major cause of morbidity and mortality with polycythemia because of the increased platelets, and blood clots can be thrown into major arteries and veins. Eventually a terminal phase of the disease has the overextended and clogged bone marrow failing and being replaced with
fibrous tissue, and in effect the system goes from overdrive to failure.

The treatment of choice for polycythemia vera is phlebotomy, or blood removal. Often a unit of blood (about 500 ml) is removed weekly until the hematocrit decreases down to 45 (the upper normal level) and is then repeated as necessary. When the disease doesn’t respond, medications called myelosuppressors are used, especially to decrease the amount of blood platelets and lessen the risk of spontaneous clotting and thrombosis. Low dose aspirin is also used as part of the regimen.

A more treatable and less serious relative is secondary polycythemia. Here, the disease is caused by conditions that increase the secretion of erythropoietin, which is a hormone produced in the kidney that stimulates the bone marrow to produce more cells. Most cases of secondary polycythemia are caused by a decrease in the amount of oxygen in the tissues (hypoxia). It can be seen in those living in high altitudes, those with extreme obesity, in smokers, and in others with chronic lung diseases.

Much of the mortality of secondary polycythemia results from a primary cause such as smoking and obesity, and leads to heart failure, angina, blood clotting and strokes due to the thrombosis caused by the elevated platelets and thick blood. In many cases, though, treatment of the primary disorder corrects the polycythemia, which then disappears as bone marrow function returns to normal.

Underwriting polycythemia vera is quite difficult; the median survival even with aggressive treatment is 10 to 15 years. Arterial thrombosis and clotting are the major causes of death, as well as leukemia due to bone marrow shutdown in about 5 percent of cases. Careful and rigorous treatment is essential in even considering a highly rated offer, and a majority of applicants may not be able to be offered life insurance coverage at all with advanced disease.

Secondary polycythemia may be insured if the primary cause is corrected and controlled—those whose cell counts remain in the upper limits of normal and who correct obvious underlying pathological conditions such as obesity, or who discontinue smoking, may only be evaluated on their primary conditions without polycythemia being a factor.