Correction: This article was corrected on September 14, 2016, to delete an incorrect statement about nonmelanoma skin cancer.

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NOTABLE NOTES

Porphyria and Vampirism-A Myth, Sensationalized

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Imagine that seated in the waiting room of your office is a pale stranger wearing a flowing black cape. In a halting Slavic accent, he complains of an aversion to sunlight and claims that his condition temporarily improves with ingestion of exogenous blood. While your instinct may be to protect your neck and run for safety, perhaps as his physician the next best step is to perform a urinalysis and hematologic testing!

Porphyria refers to a group of disorders characterized by defects in the biosynthetic pathway of heme, an erythrocyte cofactor essential for oxygen transport. Manifestations of this disease occur secondary to the accumulation of intermediates called porphyrins, which can cause photomutilation in sun-exposed areas of the skin. It was not until the 1980s, however, that porphyrias were first postulated as the inspiration for the myth of vampirism.

While dramatic and mysterious, the pale and day-fearing vampire with which we are familiar is a fairly recent creation. Prior to Bram Stoker's rendition of the creature, they were often portrayed with ruddy complexions and showed no aversion to sunlight.³ To further dispel the myth, there is no physiologic explanation as to why people with porphyria may wish to drink blood or exhibit an aversion to garlic.

Further notoriety can be attributed to a poor understanding of the dermatologic manifestations of the disease. In cutaneous porphyria, the timeframe, cause, and severity are often misunderstood. The suprabasal vesicles of porphyria cutanea tardae, the most common subtype, typically present later in life and are most often elicited by infection or hepatotoxin exposure. Less treatable porphyrias, such as erythropoietic protoporphyria, are extremely rare diseases of infancy, in which both outdoor and indoor light can lead to scarring in exposed skin.²

The dubious link between porphyria, vampirism, and the occult has had resounding effects on the perception of those with porphyria. Variegate porphyria is an autosomal dominant disease most commonly seen in South Africa. Symptoms are often acute and multisystemic, which means it is often underdiagnosed. When awareness of porphyria became transcontinental, an Afrikaans family magazine released a sensationalist article entitled "Draculas Do Indeed Exist" which stated that a "substantiated" bloodlust condition was regionally commonplace.¹ Such outrageous media led to the ostracization of many porphyria patients and deterred many others from seeking medical advice.

While the connection to vampiric myth is largely based on conjecture and misinformation, there may be little harm in keeping alive a timeold tale of mystery and intrigue as long as its connection to the debilitating disease of porphyria is severed. As the pale stranger in your office can attest, life with porphyria is difficult enough without everyone thinking you are Dracula.

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