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EMS Professionals

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TREATING

MONSTERS

Medical Causes of Monster Myths

WHAT'S THE WORST THING YOU HAVE EVER SEEN?

This question is common for medical providers once strangers learn about our daily lives. The answers can be varied, but everyone asks. I once had a child ask me during a public education event, so I turned the question on him. His answer was monsters. It brings up the time of year.

Did you have a good New Year's Eve? Not December 31, rather the one that falls on October 31, when our ancestors who lived in what are now the British Isles and northern France celebrated the end of the year. That's when the dead returned to Earth, or so they believed. Hundreds of years later, we celebrate it as Halloween.

Halloween is still a holiday surrounded by myth, mystery and tales of death. But some of the characters we have come to associate with Halloween may not be mythical mysteries, but medical mysteries. Are the bag-bearing Draculas, Wolfmen and giants simply figures developed by literature or Madison Avenue? Or are they reflections of overactive imaginations of people who lived centuries ago in a time when people suffering from various diseases were tricked instead of treated?

Monsters of folklore may not have been

created solely by the minds of storytellers, but may have had a basis in rare, but real, medical disease processes. There are many uncanny similarities between these characters and certain diseases that we see today.

Werewolves

Throughout Europe during the Medieval Age, perhaps no animal caused greater fear and alarm than the wolf. Wolves attacked livestock, were symbols of the Devil, and prevented little girls from reaching their grandmothers in fairy tales. As pack hunters, wolves were a great fear for farmers and villagers. They could mean death from the attack itself or death through starvation as wolves ravaged their

Tapetum Lucidum
Improved night vision
Yellow / amber

4 canines

12 incisors

16 premolars

Increased

bone

density



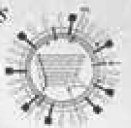
Enhanced strength
muscle mass

5 fingers

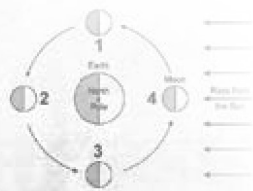
Claws -
distal phalanx

up to 6" in length

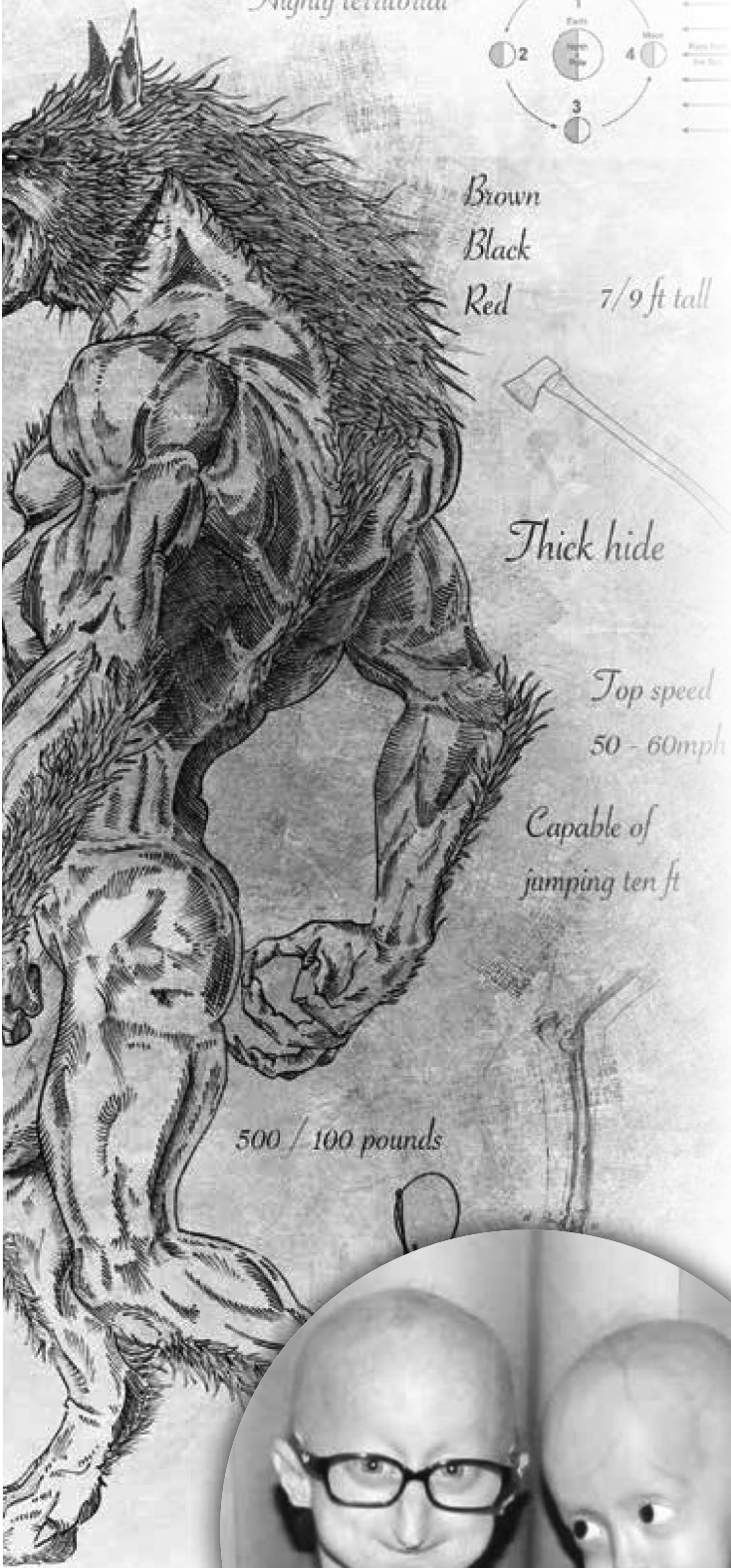
Lupine parvovirus
Pread by bite



Highly territorial



by **Christopher Suprun**, NRP, FP-C



flocks of sheep, goats and cows.

Some people took this fear one step further and had a real and significant worry that people could turn into wolves. Could a man actually become a werewolf? We know historically that many individuals died at the hands of ecclesiastic courts for the crime of being a werewolf. Evidence presented was unusually weak by our standards, and included persons who had hair growing on their palms, long fingers, or eyebrows that grew together. In some cases, though, judges appeared to recognize they might be dealing with mental conditions rather than monstrous and demonic transformations. In one case in France, a judge decided that the defendant was "stupid and idiotic," after the defendant had confessed to being a werewolf.

This recognition serves as one of two possible medical causes of the belief in werewolves. The first is a mental disease called **lycanthropy**: "The delusion of ability to transform oneself or another into an animal, especially a wolf."

In a 1977 case study for the *American Journal of Psychiatry*, two physicians outlined a case of a 49-year-old woman who described herself as a werewolf. Their report noted her demeanor to include growling, gnawing at the bed, and experiencing "auditory hallucinations." The two physicians also noted no drug involvement or alcoholic intoxication apparent in their patient.

Their treatment included neuroleptic medications, and they outlined several other disease processes that might be contributing to the patient's condition, including schizophrenia. Because of this, EMS should take all steps necessary to protect themselves and involve the appropriate law enforcement authorities as soon as possible should a possible lycanthrope present itself around Halloween.

Another disease that might have led people to believe that others were werewolves is **congenital erythropoietic porphyria**. This disease, which was theorized as early as October 1963 when Dr. Lee Illis presented a paper about the mysterious werewolf disease that howled through Europe in the Middle Ages, is the development of a recessive gene that lacks a specific enzyme, called **uroporphyrinogen III cosynthase**.

Signs and symptoms described for congenital erythropoietic porphyria include extreme photosensitivity which may lead to blistering, severe scarring and increased hair growth. The patient will likely show signs and symptoms of abdominal pain, urine that is pink or dark brown, and



disorientation, confusion and psychotic behavior. Additionally, the patient may have erythrodontia or a reddening of the teeth from porphyrins being deposited into the tooth. These symptoms might cause a person suffering from this disease to walk hunched over and with the look of blood on their mouth as if they had recently devoured their latest victim.

With the extreme agitation that is sometimes associated with the disease, the approach by others might further agitate the patient.

Vampires

Another disease we need to investigate when considering monster folklore is a hereditary condition called xeroderma pigmentosum, or XP, which is caused by a recessive gene. With XP, the skin is normal at birth, but has an "extreme sensitivity to sunlight." XP is a disease in which skin cells cannot repair DNA damage as normal cells would, so exposure to ultraviolet light (including sunshine), can cause permanently damaged cellular tissue. In the U.S., XP occurs about once every 250,000 births and starts in infancy with erythema, or red spots, on sun-exposed areas of the face, hands and neck. Freckling and scaling follow, along with skin atrophy and the development of benign and malignant tumors.

It is important to note also that the patient is not suffering from albinism, which is a disease involving the lack of pigment in the skin. Persons suffering from XP have a cellular DNA replication defect at the cellular level, whereas patients with albinism lack

pigment, but their DNA repair structure works correctly.

Finally, XP is often joined by photophobia and conjunctivitis in about 80% of cases.

One monster from folklore that probably was created from some semblance of reality and perhaps this disease process is Dracula. The vampire Dracula, famous from Bram Stoker's 1897 novel, was most likely inspired by a Transylvanian prince named Vlad IV. Vlad, whose nickname or surname was Dracula, meant "dragon" or "devil" to his detractors. This nickname came from his induction into a pseudo-religious military order called the Order of the Dragon. A characteristic of this Order was its use of "a black cape over a red garment," very consistent with our image of vampires.

Vlad Dracula was a merciless tyrant. Though he had numerous methods of torture and execution for men, women and children, he emphasized a particularly ruthless method of death: impalement.

Vlad the Impaler, as he is also known, would impale his victims through their entire body and is said to have impaled as many as 20,000 Turkish captives in an effort to terrorize attackers. During one campaign against him, he poisoned wells and burned his own villages, helping to create the bloodthirsty nature of the Dracula that we associate with the vampire today.

There are several instances where the myth and the man come together. A ruthless "taste for blood" is an obvious allusion, given his proclivity

for killing. The black cape and red garments add to the picture, and there is a certain irony to his use of impaling stakes, likely wooden, that throughout literature are used to kill vampires.

Villagers in and around the countryside reported Vlad Dracula was also known to have "roamed across the countryside in disguise, particularly at night." Was this because he was a vampire, or was it because he suffered from the photophobias and conjunctivitis associated with XP? This fear or sensitivity to sunlight rings true when we consider not only the myths that surround the vampire tradition and having red, or "pink eye" such as conjunctivitis, but the medical issues surrounding XP and its associated diseases. All those things together lend themselves to a bad cliché: They add just "one more nail in the coffin" in a case that medieval "vampires" were really patients in need of a good dermatologist; they were suffering from XP.

There is little an EMS provider can do to treat these patients, but should you encounter one of these patients, remember to avoid UV light as much as possible. Most of these patients will be aware of their condition and may have special body suits for moving around outside. If you do transport a patient with XP, special caution should be given to covering windows during the transport and preventing light from entering the ambulance.

Giants and Little People

An EMS case study on giants might include a patient who is

significantly taller than average, nearing 7 or 7.5 feet. On examination, the patient might have an obvious overbite with spaced teeth, a very deep voice and speak in garbled words, and thick leathery skin. The patient might complain of tingling in the hands or stiffness in his joints.

This "monster" we have heard about since childhood is the giant. Giants can be found everywhere in children's stories, from the Bible's Goliath to Grimm's "Jack and the Beanstalk" to William Steig and Dreamworks' "Shrek." Whether or not these childhood cases were giants or if they suffered from a medical condition will probably never be known, but two medical conditions that can cause this excessive growth do exist.

Gigantism and acromegaly, are caused by excessive exposure to growth hormone (GH). When this occurs prior to epiphyseal plate fusion, a youth is diagnosed with gigantism. If the bone has ceased its growth, it is acromegaly. Adenomas, or benign tumors that are affixed to the pituitary gland, are responsible for 90% of acromegaly cases.

The world's tallest man, Robert Wadlow, attained a height of 8 feet, 11 inches. He died at age 22, weighing 490 pounds. To give a proper sense of his development, he weighed 8 pounds, 6 ounces at birth. At six months, he weighed 30 pounds, and at age 8, he was 6 feet 2 inches tall and weighed almost 200 pounds.

These patients suffer significant overgrowths of tissue. As it thickens, the tissue can trap nerves, causing numbness and weakness in the

hands. Enlarged sinus cavities lead to deep voices. Diabetes, hypertension and cardiac hypertrophy should be expected by EMS providers dispatched to giants, or persons with a history of acromegaly.

These patients can be treated by irradiation or surgical removal of the tumor. Additionally, bromocriptine, which has been used to treat Parkinson's Disease and forms of infertility in males and females, has had some success blocking the release of GH in patients with acromegaly and gigantism.

Somavert, currently in clinical trials, controls the effects of excess GH by acting as a growth hormone receptor, interfering with normal GH cellular binding. This drug may be years away from availability.

Robert Wadlow died in 1940, well before imaging of the pituitary gland was available. EMS will need to focus on individualized supportive care for their response to neighborhood giants.

On the opposite end of the spectrum, victims of Hutchinson-Gilford Syndrome bear a burden as great as any of the other patients cited. This disease, also known as progeria, tears life from the young through what has recently been found to be a DNA obstruction. The disease prematurely ages individuals afflicted with it and has an incidence of about one case per 4 million to 8 million births.

Patients diagnosed with progeria often have normal births and infancy through their first year, but their



growth is quickly stunted, usually by 18 to 24 months. Signs and symptoms of progeria include profound growth failure, loss of body fat and hair, and aged looking skin. These children die of cardiovascular disease, primarily atherosclerotic buildups, between ages 8 and 21. The average life expectancy for a child diagnosed with progeria is 13.

Progeria's cause was first reported in April 2003 as a "mutation in the Lamin A" protein. "Lamin A protein is the structural scaffolding that holds the nucleus together and is involved in gene expression and DNA replication," according to the Progeria Research Foundation's website.

Unfortunately, EMS' ability to render care to these children will be primarily focused on standard ACLS approaches to cardiovascular conditions, preferably administering weight-dosed drugs.

Treats Instead of Tricks

As you recall the ghosts and goblins that knock at your door, remember that some of these costumes can be traced back to persons who were attacked as evil or demonic beings, but who may have suffered through extraordinarily rare and debilitating diseases.

Perhaps more treats for our little representatives of past myths will keep

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the hospital is on diversion, the hospital has an EMTALA duty to that patient.

- CMS considers the “parking” of patients in hospitals and refusing to release EMS equipment or personnel as jeopardizing patient health while impacting the ability of the EMS personnel to provide emergency services to the rest of the community. Depending upon the facts of the case, the practice could constitute an EMTALA violation. 🌀

Gregory West is a licensed attorney in Wisconsin and has more than 27 years of experience as an EMS provider and firefighter. Gregory is the Dean of Protective and Human Services at Waukesha County Technical College. He teaches law to emergency providers on a part-time basis and also serves as a member of the State EMS Board.

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TREATING MONSTERS

our focus on treating the diseases for which so many have and do suffer innumerable tricks. 🌀

Christopher Suprun is a 25-plus year firefighter/paramedic. He has spoken across the U.S. and internationally, and is Director of Education and Outreach for the Never Forget Foundation.

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