

In the medical community, the term “zebra” is universally understood as a reference to an orphan, or rare disease. Physicians are taught the core tenet of medical diagnosis—to assume that the simplest explanation is usually the best, i.e., it is generally more productive to look for common rather than exotic causes for disease, hence the phrase “if you hear hoofbeats, think horses, not zebras.” This is a modern day variant of Occam’s Razor which states “one should not make more assumptions than needed; when multiple explanations are available—choose the simplest. **Carcinoid and NETS** are zebra diseases...

it’s all about the stripes!

MISSION DVCC

Creating awareness and a bridge to resources, support, education and the medical community for Carcinoid cancer & neuroendocrine tumor patients and their advocates.

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Zebra illustration donated by
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*It's All
About
The Stripes*



HOOFBEATS

The DVCC has adopted a zebra through the Philadelphia Zoo’s adopt-an-animal program. The zoo is happy to offer its support of this effort, which the DVCC calls “Hoofbeats” since this rare cancer has the zebra as its icon. The Carcinoid community thanks Dr. Eugene Woltering, of the *Neuroendocrine Clinic* at Ochsner Medical Center (in affiliation with Louisiana State University Health Science Center) in Kenner Louisiana, for sharing his marketing acumen, devotion, and enthusiasm for all things striped, especially humans.

Since our mission includes creating awareness of the disease in both the medical and patient communities, the consensus was there was no better, no more interesting way to make a statement. The Philadelphia Zoo is the first zoo to join this awareness campaign.

INFORMATION RESOURCES

www.carcinoid.org— website of the Carcinoid Foundation. Since 1968 this organization has been dedicated to educating patients and medical professionals about carcinoid and neuroendocrine tumors (NETs) and encouraging and supporting research aimed at improving diagnosis, treatment, and searching for a cure.

www.carcinoid.org/pcf/lectures/index.shtml—webcast lectures

www.ochsner.org/page.cfm?id=1129 The neuroendocrine program at Ochsner Medical Center in association with Louisiana State University Health Center.

www.us.sandostatin.com—Novartis Pharmaceuticals

www.pharma.us.novartis.com—Novartis-main site

www.interscienceinstitute.com—publisher of Neuroendocrine Tumors: A comprehensive Guide to Diagnosis and Management

www.ccan.org Carcinoid Cancer Awareness Network

www.caringforcarcinoid.org— foundation supporting research and dedicated to finding a cure

www.cancer.gov National Cancer Institute

<http://www.nlm.nih.gov/medlineplus/carcinoidtumors.html>

US National Library of Medicine & National Institutes of Health

clinicaltrials.gov/search/open/condition=%22Carcinoid+Tumor%22—current clinical trials

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What is Carcinoid Cancer?

The most common type of neuroendocrine tumor (NET), Carcinoid, has been called "cancer in slow motion" because typically they tend to grow so slowly that people afflicted with these tumors usually live for many years, sometimes for a normal life span. The less common, atypical Carcinoid, is faster-growing and aggressive. However, the wide variety of new treatments make the outlook for most atypical victims more hopeful than before.

A neuroendocrine (NET) tumor originates from the hormone-producing cells of the body's neuroendocrine system, which consists of cells that are a cross between traditional endocrine cells (or hormone-producing cells) and nerve cells. Neuroendocrine cells are found in organs such as the lungs and gastrointestinal tract, however, Carcinoid can appear in different areas of the body besides the gastrointestinal tract and the lungs, including the ovaries, testicles, urinary bladder, the prostate, the breast, kidneys, thymus, and very rarely the eye and ear. Besides the physicality of the tumors,

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the secretion of excessive hormones such as serotonin affect the functioning of other systems of the human body. These symptoms vary according to the type and amount of hormone, location of the tumor and if the cancer has metastasized. Carcinoid Syndrome is one manifestation of excess hormones and usually presents with flushing and diarrhea. Carcinoid is both misdiagnosed and under diagnosed. The disease can masquerade as irritable bowel syndrome, Crohn's disease, asthma, menopause, rosacea, or even acid reflux - **which is a reason that it can be many years before a diagnosis is made.**



If you don't suspect it, you can't detect it!

N.E.T.

About 11,000 to 12,000 neuroendocrine tumors (NETs) and cancers are diagnosed each year in the United States. 2 out of 3 of these tumors occur in the digestive system. They are the most common NETs of the gastrointestinal tract.



How is it diagnosed?

Once Carcinoid is suspected, the diagnosis usually can be confirmed quickly by doing a 24 hour urine 5-HIAA test (the main waste product of serotonin). In the presence of Carcinoid Syndrome the amount of 5-HIAA is almost always distinctly above normal. Certain foods and medicines must be avoided for a day or two before and the day of the urine collection since they can cause false test results. In 1/3 of Carcinoids this test is negative, therefore both the 5-HIAA and a chromogranin A (CgA) should initially be done together, possibly with a serotonin blood level. The measurement of CgA is considered "the gold standard" of chemical tests for confirming the diagnosis of carcinoid and NETs and for following their course.

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The first diagnostic study recommended is the "gold standard" for finding Carcinoid/NET tumors - the **OctreoScan**. It is successful in 85% of carcinoids and consists of a harmless injection of a minute dose of a short duration radioactive isotope specifically attracted to carcinoid/NET tumor tissue which then lights up when a radiation scan is taken over the entire body. An OctreoScan is recommended in almost all cases even when the diagnosis is known. This is very important in cases where standard imaging and chemical markers fail to reveal the diagnosis/location of tumors. There are some cases in which all the symptoms and chemical findings of Carcinoid Syndrome are present but standard tests fail to reveal a tumor. In these cases, an octreoscan can be a great help in confirming the diagnosis and locating the tumor(s). A positive octreoscan usually predicts a good response to treatment with octreotide (Sandostatin) and is necessary for certain systemic radioisotope treatments (Lu-177, Yttrium 90). Other supplementary imaging techniques are chest X-rays, CT scans, MRIs, upper GI, small bowel X-ray studies, FDG or F18 Dopa PET scans, MIBG scans.

How is it treated?

Surgery, with complete removal of the primary tumor tissue, is the first and best treatment when possible, and **if detected early, can result in a complete and permanent cure.** Somatostatin analogue injections not only usually relieve symptoms of Carcinoid Syndrome but are now believed to sometimes inhibit or even reverse tumor growth. This has become the main treatment for most, with or without Carcinoid Syndrome. When it has metastasized to the liver, radiofrequency ablation (RFA) is one treatment now used to destroy the metastases when it has not been possible to excise them surgically. Another way to debulk unresectable Carcinoid liver tumors is to inject the liver artery supplying blood to the metastases with a combination of embolic material and chemotherapy drugs. This shuts off the blood flow to the tumors and also loads them with tumor destroying/growth inhibiting chemotherapy. Thus this chemotherapy is concentrated in the tumors where it can have a much greater effect. Another new treatment for liver metastases consists of injecting the hepatic artery with microspheres filled with radioactive isotopes.

A number of combinations of chemotherapy drugs have shown to be beneficial, however, the site of the primary tumor influences the tumors' response to chemotherapy. Any treatment should be individualized for each patient.

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This information is extrapolated from www.carcinoid.org, website of the Carcinoid Foundation, Inc.—(a federally chartered not-for-profit organization). The documentation was written by Dr. Richard R. P. Warner who is an internationally known expert in Carcinoid and NETs and a pioneer in the field for 50 years. It's good idea to make this website your first stop for knowledge about this disease.

