NAVIGATING THE WORLD OF NEUROENDOCRINE TUMORS

for patients and their primary care physicians

zebra talk

Introduction

Welcome to the second edition of *Zebra Talk* . We think we've produced an awesome handbook that's both practical and has universal appeal. It is not intended as medical advice, but rather a collection of useful facts and pursuable options which we hope will inspire thoughtful communication between patients, their physicians and caregivers.

Zebra Talk can help level out the playing field of informed decision-making for your medical care. Sometimes information won't pertain to you; but, at other times, it will be spot on. We think we've chosen wisely— our editorial staff volunteers either have a disease on the NET spectrum, or work in the medical field. Dr. Eugene Woltering, Clinical Director of the NOLANETs Clinic in New Orleans, Louisiana, is our honorary medical editor who looks over our shoulder so we won't dot t's or cross i's. His guidance ensures this project's integrity and we are humbly grateful for his participation.

Zebra Talk is not the be all, end all of information about NET diagnosis, characteristics, treatments and resources, but it's a platform of a wide variety of practical information mixed with a dollop of experience and, this year, we've included a generous amount of research links for online exploration. *Our mission is to help you become knowledgeable about the hows and whys and the management of your disease.* We hope you'll find success in treating your disease effectively and living your life with — and in spite of— the quirky and mysterious world of NETs. We are the odd men out—a rare cancer basically no one has heard of. But we've made progress in the last ten years and have actually raised the bar for rare cancer awareness in many ways. In most cases, the complexities of neuroendocrine tumors are daunting to both the patients and their primary care physicians, who very rarely, if ever, see a case. Optimally, health decisions should be made in partnership with your physician (s) and quite often there is a knowledge deficient on both sides. Today, most NETs are considered chronic diseases and we hope that by providing insights, tools, and resources we'll help to give you a leg up on living a balanced life.

This handbook has more sophisticated (*read medi-speak*) language than previous editions. To compensate, we expanded the glossary and placed many definitions within the narrative. The reason is that when we attend conferences that host NET specialists, it has become common practice for them to speak to our intellects and experience. As a result, their vocabulary is beyond that of most recently diagnosed NET patients.

Besides those in the diagnostic process, or who are recently diagnosed, *Zebra Talk* is relevant for anyone who otherwise needs to explore this bewildering array of acronyms, biochemistry, genetics and complicated exceptions to exceptions. It is our pleasure to participate in NET community awareness and efforts and to contribute to the much-needed dissemination of NET knowledge.

table of CONTENTS

LIST OF TABLES

LIST OF ILLUSTRATIONS

**Throughout Zebra Talk, neuroendocrine tumors are referred to as NETs.*

Lab Tests for NETs | Based on NOLANET's Protocol[1]

Basic Lab Workup

- CBC with diff and platelet count (complete blood count)
- CMP (complete metabolic profile)
- PT, PTT, INR
- Free T4, Free T3 and TSH
- 25-Hydroxy Vitamin D2,D3 (Dx:579.4)

Carcinoid Labs

Send to Inter Science Institute (ISI) (Only as noted). Call ISI to obtain Z tubes. 800.255.2873. Caveat: Do not use Cambridge Biomedical Lab. ISI specializes in neuroendocrine research and development; the top NET specialists and researchers are on its Medical Council.

- 5-HIAA plasma Send to ISI. NOLANETs no longer uses the 5-HIAA urine assay. Discuss with your physician.
- Chromogranin A (CgA)
- Serotonin (frozen serum)
- Pancreastatin 3CC frozen Z-tube Send to ISI
- Neurokinin A 3CC frozen Z-tube Send to ISI
- Substance P-2 CC frozen lavender top or Z-tube Send to ISI
- Neuron Specific Enolase (NSE)
- Octreotide/Sandostatin Drug Level-3CC plasma or frozen red top—Send to ISI ONLY IF Patient is on Sandostatin-Draw immediately before the next dose of Sandostatin
- Lanreotide/Somatuline depot Drug Level –3CCplasma or frozen red top-Send to ISI ONLY IF Patient is on Somatuline depot-Draw immediately before next dose of Somatuline

Gastric Carcinoid Labs

- Chromogranin A
- Anti-parietal cell AB
- Gastrin
- B-12
- Folate
- Pancreastatin
- Anti-thyroid AB
- Anti-islet cell AB
- Serotonin (serum)

Pancreatic Islet Cell Labs

- Chromogranin A
- Pancreastatin
- Gastrin
- Glucagon
- Pancreatic polypeptide
- Vasoactive intestinal polypeptide (VIP)
- C peptide
- Insulin
- Somatostatin

Other/Optional Labs

- Prolactin
- Motilin
- Ghrelin
- PTH intact with calcium
- IGF-1 (insulin-like growth factor)
- Calcitonin
- Growth Hormone
- ACTH

Flushing Work Up (782.62)

- Chromogranin A
- Pancreastatin
- Substance P
- VIP
- Gastrin
- Serotonin
- Neurotensin
- Calcitonin
- Histamine
- 24 hr. urine for 5-HIAA, VMA, histamine
- Tryptase

Diarrhea Work Up (787.91)

- Chromogranin A
- Pancreastatin
- Gastrin
- Gastrin-releasing peptide
- Calcitonin
- Histamine
- Pancreatic polypeptide
- PTH intact
- FSH
- VIP
- 24 hr. Urine for 5-HIAA, VMA

Pheochromocytoma (194.0)

- Plasma-free metanephrine & normetanephrine, plasma catecholamines (epinephrine, norepinephrine, dopamine)
- 24 hour urine for metanephrine, VMA normetanephrine
- Chromogranin A
- Pancreastatin

Table 4

Radiation Table for Common Scans

Radiation is of concern in the NET community since our diseases are chronic and followed using nuclear scans at certain intervals. The average dose of radiation per person from all sources (*not including scans*) is about **620 mrems per year.** International standards allow exposure to as much as 5,000 mrems a year for those who work with and around radioactive material. There is a downloadable copy of a complete workup chart on our website to compute your annual radiation exposure **www.phillynets.org**.

There are other measurements for radiation exposure such as an internationally recognized unit called millisieverts (mSv). based on the metric system. The conversion factor is:

100 mrems = 1 mSv

To find out your true exposure, calculate your scan exposure and add in factors such as where you live, the altitude of where you live, nearby nuclear power plants, food, water and air and factors like plane travel, computer and TV screens (CRT technology), cigarettes (1/2 pack daily for a year = 18 mrem), porcelain tooth crowns, airport x-rays, smoke detectors.[1]

Table 2

SCAN	RADIATION UNITS*
CT=computerized axial tomography scan (CAT scan)	
CT Scans Head	200 mrem
CT Scans Chest	700 mrem
CT Scans Abdomen/pelvis	1000 mrem
CT Scans Extremity	10 mrem
CT Scans Angiography (heart)	2000 mrem
CT Scans Angiography (head)	500 mrem
CT Scans Spine	1000 mrem
CT Scans Whole Body	1000 mrem
CT Scans Cardiac	2000 mrem
X-Ray Chest	10 mrem
X-Ray Mammography (breast)	42 mrem
X-Ray Skull	10 mrem
X-Ray Cervical spine	20 mrem
X-Ray Lumbar spine	600 mrem
X-Ray Upper GI	600 mrem
X-Ray Abdomen (kidney/bladder)	700 mrem
X-Ray Barium enema	800 mrem
X-Ray Pelvis	60 mrem
X-Ray Dental bitewing/image	.5 mrem
X-Ray Hip	70 mrem
X-Ray Extremity (hand/foot)	.5 mrem
DEXA Scan (bone mineral density)	2 mrem

Chart from the American Nuclear Society

* REM is the acronym for Roentgen Equivalent Man. A millirem (mrem) is one thousandth of a rem , i.e., 1000 mrem = 1 rem. Mrem is a very small measurement.

NETbits

The **OctreoScan™**, which is the gold standard diagnostic scan for NETs, has a dose of radiation measuring 2600 mrems. If a SPECT/CT scan is added to the procedure you can receive an additional 400-700 mrems according to the technique.

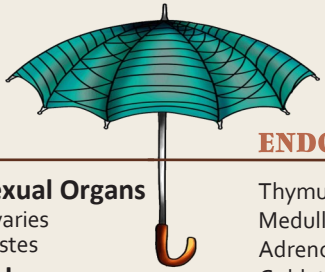
The ^{68}**Gallium DOTATOC scans** which are in clinical trial in the United States, result in a dose of about 450 mrems. The radiation from a CT scan would add another 400-700 mrems. The overall dose to a patient is much lower for the ^{68}gallium DOTATOC scan than for the OctreoScan™.

Types of NETs

At one time carcinoid was the term used for all NETs; it is now common practice to recognize it as a sub-group of the umbrella group neuroendocrine tumors (NETs). G.I. NETs (gastrointestinal) and P-NETs (pancreatic) are often grouped together as **GEP NETs**—**G**astro**E**ntero**P**ancreatic **N**euro**E**ndocrine **T**umors.

Neuroendocrine Tumors

Table 1



CARCINOID

GI Carcinoids
Stomach (gastric)
Duodenum
Jejunum
Ileum
Right Colon
Transverse Colon
Rectum
Liver (metastatic)
Gall Bladder
Cecum

Goblet Cell
Appendix

Sexual Organs
Ovaries
Testes

Pulmonary
Atypical
Typical
Esophageal

Small Cell Lung Carcinoma

Large Cell Neuroendocrine Carcinoma

ENDOCRINE NETS

Thymus Neuroendocrine Cancer
Medullary Thyroid Carcinoma
Adrenocortical Carcinoma
Goblet Cell
Neuroblastoma
Merkel Cell Carcinoma
Paragangliomas &
Pheochromocytomas

NEUROTENSINOMAS

UNKNOWN PRIMARY

PANCREATIC NETs

Gastrinomas
VIPomas
ACTHoma
Insulomas
Glucaonomas
Polypeptidomas
Somatostatinomas

FAMILIAL SYNDROMES

MEN 1
MEN2A
MEN2B

Why the zebra?

In the medical community, the term **"zebra"** is a universal reference to an **orphan** (*rare*) disease. Physicians learn the core tenet of diagnosis—to assume that the simplest explanation is usually the best, so, it is generally more productive to look for common, rather than exotic causes for diseases, hence the phrase, *"If you hear hoof beats, it's probably horses, not zebras."* (Based on *Occam's Razor* principle — i.e., the simplest, most logical answer is often the correct one). To encourage disease recognition, knowledge and awareness, the worldwide NET community has adopted the zebra to symbolize our uniqueness and create awareness through the zebra's eye-catching appeal of its flamboyant camouflage (the editor's favorite oxymoron).

WORLDWIDE NET CANCER AWARENESS DAY

"The International Neuroendocrine Cancer Alliance (INCA) is the global voice in support of neuroendocrine cancer patients. We constantly need to raise awareness of NET cancers among decision makers, health professionals and the general public. Information is key to improving quality of life and prognosis for NET cancer patients; raising awareness is therefore one of our primary goals."

HELD ANNUALLY ON NOVEMBER 10
WWW.NETCANCERDAY.ORG

The information and resources contained in Zebra Talk are useful for sourcing information, but are not substitutes for professional medical advice, care or any recommended treatments by your specialist and/or your primary care physician.

Publisher: The Patients' Project, Inc. A federal 501 (c) 3 not-for-profit.
821 Meadow Drive, Warminster PA 18974 USA

MANY THANKS

to our sponsors
Ipsen Biopharmaceuticals
Pfizer Inc., BerriHealth, and
InterScience Institute for
contributing their generous
support for this project.

Specializing in NETs laboratory testing:
www.interscienceinstitute.com
InterScience Institute

IPSEN
Innovation for patient care

berrihealth

Pfizer

A carry along resource handbook for NET patients and their primary care physicians. We hope this handbook will inspire communication between patients, their physicians, and their caregivers.