

Consensus Statements and Guidelines

This section provides information on consensus findings from leading international and national researchers and physicians in the field of carcinoid and neuroendocrine tumors.

The North American Neuroendocrine Tumor Society (NANETS) Guidelines

The publication of the first **North American consensus guidelines for NET disease management** was a major achievement for the carcinoid/NET community in 2010. These guidelines include early detection procedures for diagnostic purposes, imaging, histopathology, biochemical evaluation, surgical procedures, and evidence-based treatments emphasizing a multidisciplinary team-based approach to patient care. The NANETS Guidelines Working Group consists of 38 carcinoid/NET specialists writing guidelines in the areas of Epidemiology and Genetics, Pathology, Radionuclide Therapies, Biochemistry, Symptomatic Control, Surgery, Imaging and Liver-Directed Therapies, Systemic Treatments, and Follow-up. The guidelines have been published in **Pancreas**, Volume 39, Number 6, August 2010. They are as follows:

[The North American Neuroendocrine Tumor Society \(NANETS\) Guidelines: Mission, Goals, and Process.](#)

Authors: Kvolts, Larry K. MD *; Brendtro, Kari L. +

DOI: DOI: 10.1097/MPA.0b013e3181eb7451

[The Pathologic Classification of Neuroendocrine Tumors: A Review of Nomenclature, Grading, and Staging Systems.](#)

Authors: Klimstra, David S. MD *; Modlin, Irvin R. MD, PhD +; Coppola, Domenico MD ++; Lloyd, Ricardo V. MD, PhD [S]; Suster, Saul MD [//]

DOI: DOI: 10.1097/MPA.0b013e3181ec124e

[NANETS Consensus Guidelines for the Diagnosis of Neuroendocrine Tumor.](#)

Authors: Vinik, Aaron I. MD, PhD *; Woltering, Eugene A. MD +; Warner, Richard R. P. MD ++; Caplin, Martyn MD [S]; O'Dorisio, Thomas M. MD [//]; Wiseman, Gregory A. MD [P]; Coppola, Domenico MD #; Go, Vay Liang W. MD **

DOI: DOI: 10.1097/MPA.0b013e3181ebaffd

[NANETS Treatment Guidelines: Well-Differentiated Neuroendocrine Tumors of the Stomach and Pancreas.](#)

Authors: Kulke, Matthew H. MD *; Anthony, Lowell B. MD +; Bushnell, David L. MD ++; de Herder, Wouter W. MD, PhD [S]; Goldsmith, Stanley J. MD [//]; Klimstra, David S. MD [P]; Marx, Stephen J. MD #; Pasiaka, Janice L. MD **; Pommier, Rodney F. MD ++; Yao, James C. MD +++; Jensen, Robert T. MD [S][S]

DOI: DOI: 10.1097/MPA.0b013e3181ebb168

[The NANETS Consensus Guideline for the Diagnosis and Management of Neuroendocrine Tumors: Well-Differentiated Neuroendocrine Tumors of the Jejunum, Ileum, Appendix, and Cecum.](#)

Authors: Boudreaux, J. Philip MD *; Klimstra, David S. MD +; Hassan, Manal M. MD, PhD ++; Woltering, Eugene A. MD *; Jensen, Robert T. MD [S]; Goldsmith, Stanley J. MD [//]; Nutting, Charles DO [P]; Bushnell, David L. MD #; Caplin, Martyn E. MD **; Yao, James C. MD ++

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Published on The Carcinoid Cancer Foundation (<http://www.carcinoid.org>)

DOI: DOI: 10.1097/MPA.0b013e3181ebb2a5

[The NANETS Consensus Guidelines for the Diagnosis and Management of Gastrointestinal Neuroendocrine Tumors \(NETs\): Well-Differentiated NETs of the Distal Colon and Rectum.](#)

Authors: Anthony, Lowell B. MD *; Strosberg, Jonathan R. MD +; Klimstra, David S. MD ++; Maples, William J. MD [S]; O'Dorisio, Thomas M. MD [//]; Warner, Richard R.P. MD [P]; Wiseman, Gregory A. MD #; Benson, Al B. III MD **; Pommier, Rodney F. MD ++

DOI: DOI: 10.1097/MPA.0b013e3181ec1261

[The North American Neuroendocrine Tumor Society Consensus Guideline for the Diagnosis and Management of Neuroendocrine Tumors: Pheochromocytoma, Paraganglioma, and Medullary Thyroid Cancer.](#)

Authors: Chen, Herbert MD *; Sippel, Rebecca S. MD *; O'Dorisio, M. Sue MD, PhD +; Vinik, Aaron I. MD, PhD ++; Lloyd, Ricardo V. MD, PhD [S]; Pacak, Karel MD, PhD, DSc [//]

DOI: DOI: 10.1097/MPA.0b013e3181ebb4f0

[NANETS Consensus Guideline for the Diagnosis and Management of Neuroendocrine Tumors: Well-Differentiated Neuroendocrine Tumors of the Thorax \(Includes Lung and Thymus\).](#)

Authors: Phan, Alexandria T. MD *; Oberg, Kjell MD, PhD +; Choi, Junsung MD ++; Harrison, Lynn H. Jr MD [S]; Hassan, Manal M. MD, PhD [//]; Strosberg, Jonathan R. MD [P]; Krenning, Eric P. MD, PhD #; Kocha, Walter MD **; Woltering, Eugene A. MD ++; Maples, William J. MD ++++

DOI: DOI: 10.1097/MPA.0b013e3181ec1380

[The NANETS Consensus Guidelines for the Diagnosis and Management of Poorly Differentiated \(High-Grade\) Extrapulmonary Neuroendocrine Carcinomas.](#)

Authors: Strosberg, Jonathan R. MD *; Coppola, Domenico MD +; Klimstra, David S. MD ++; Phan, Alexandria T. MD [S]; Kulke, Matthew H. MD [//]; Wiseman, Gregory A. MD [P]; Kvols, Larry K. MD *

DOI: DOI: 10.1097/MPA.0b013e3181ebb56f

[Consensus guidelines for the management and treatment of neuroendocrine tumors.](#)

Pancreas. 2013 May;42(4):557-77.

Authors: Kunz PL1, Reidy-Lagunes D, Anthony LB, Bertino EM, Brendtro K, Chan JA, Chen H, Jensen RT, Kim MK, Klimstra DS, Kulke MH, Liu EH, Metz DC, Phan AT, Sippel RS, Strosberg JR, Yao JC; North American Neuroendocrine Tumor Society. **DOI:**10.1097/MPA.0b013e31828e34a4

European Neuroendocrine Tumor Society (ENETS) 2012 Guidelines, TNM Grading, Standards of Care, and Metastases

- [Introduction](#)
- [Gastroduodenal Neoplasms](#)
- [Colorectal Neuroendocrine Neoplasms](#)
- [Functional Pancreatic Endocrine Tumor Syndromes](#)
- [Well-Differentiated Pancreatic Non-Functioning Tumors](#)
- [Neuroendocrine Neoplasms from the Jejunum-Ileum and the Appendix, including Goblet Cell Carcinomas](#)
- [Liver and Other Distant Metastases from Neuroendocrine Neoplasms of Foregut, Midgut.](#)

[Hindgut and Unknown Primary](#)

ENETS 2011 Consensus Guidelines

- [ENETS 2011 Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Tumors: An Update](#)
- [ENETS Consensus Guidelines for the Management of Patients with Gastroduodenal Neoplasms](#)
- [ENETS Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Neoplasms: Colorectal Neuroendocrine Neoplasms](#)
- [ENETS Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Neoplasms: Functional Pancreatic Endocrine Tumor Syndromes](#)
- [ENETS Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Neoplasms of the Digestive System: Well-Differentiated Pancreatic Non-Functioning Tumors](#)
- [ENETS Consensus Guidelines for the Management of Patients with Neuroendocrine Neoplasms from the Jejunum-Ileum and the Appendix Including Goblet Cell Carcinomas](#)
- [ENETS Consensus Guidelines for the Management of Patients with Liver and Other Distant Metastases from Neuroendocrine Neoplasms of Foregut, Midgut, Hindgut, and Unknown Primary](#)

UK and Ireland Neuroendocrine Tumour Society (UKI NETS) Guidelines

- [Guidelines for the management of gastroenteropancreatic neuroendocrine \(including carcinoid\) tumours \(NETs\)](#)
Gut. 2012 Jan;61(1):6-32. doi: 10.1136/gutjnl-2011-300831. Epub 2011 Nov 3.
Ramage JK, Ahmed A, Ardill J, Bax N, Breen DJ, Caplin ME, Corrie P, Davar J, Davies AH, Lewington V, Meyer T, Newell-Price J, Poston G, Reed N, Rockall A, Steward W, Thakker RV, Toubanakis C, Valle J, Verbeke C, Grossman AB; UK and Ireland Neuroendocrine Tumour Society.

Abstract: These guidelines update previous guidance published in 2005. They have been revised by a group who are members of the UK and Ireland Neuroendocrine Tumour Society with endorsement from the clinical committees of the British Society of Gastroenterology, the Society for Endocrinology, the Association of Surgeons of Great Britain and Ireland (and its Surgical Specialty Associations), the British Society of Gastrointestinal and Abdominal Radiology and others. The authorship represents leaders of the various groups in the UK and Ireland Neuroendocrine Tumour Society, but a large amount of work has been carried out by other specialists, many of whom attended a guidelines conference in May 2009. We have attempted to represent this work in the acknowledgements section. Over the past few years, there have been advances in the management of neuroendocrine tumours, which have included clearer characterisation, more specific and therapeutically relevant diagnosis, and improved treatments. However, there remain few randomised trials in the field and the disease is uncommon, hence all evidence must be considered weak in comparison with other more common cancers.

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Published on The Carcinoid Cancer Foundation (<http://www.carcinoid.org>)

PMID: 22052063 [PubMed - indexed for MEDLINE]

PMCID: PMC3280861

[Read](#) full text.

- [Nordic Guidelines 2010 for the Diagnosis and Treatment of Gastroenteropancreatic Neuroendocrine Tumors](#)
(click on the title for full PDF article)
- [Consensus recommendations for the diagnosis and management of well differentiated gastroenterohepatic neuroendocrine tumours: a revised statement from a Canadian National Expert Group \(2010\)](#) (click on the title for full PDF article)
- [ENETS Consensus Guidelines for the Diagnosis and Treatment of Neuroendocrine Gastrointestinal Tumors](#)

Part 2 - Midgut and Hindgut Tumors

Published in Neuroendocrinology Special Issue: 2008, Vol. 87, No. 1

Editor(s): [de Herder, W.W.](#) (Rotterdam); [O'Toole, D.](#) (Dublin); [Rindi, G.](#) (Parma); [Wiedenmann, B.](#) (Berlin) Click on each Editors name and get additional neuroendocrine related articles.

Readers will be able to find answers to debated questions for specific tumors, such as the diagnostic procedure for midgut classic carcinoids, follow-up for appendix carcinoids, or the algorithm for liver metastasis treatment of patients with extrahepatic spread from gut neuroendocrine carcinomas.

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- [ENETS \(European NeuroEndocrine Tumor Society\)](#)

Become a member and get access to the most comprehensive guidelines.

- Recommendations for radioembolization of hepatic malignancies using Yttrium-90 microsphere brachytherapy.

[A consensus panel report from the radioembolization brachytherapy oncology consortium](#) (Full text PDF)

Int. J. Radiation Oncology Biol. Phys., Vol. 68, No. 1, pp. 13-23, 2007

PURPOSE: To standardize the indications, techniques, multimodality treatment approaches, and dosimetry to be used for yttrium-90 (Y90) microsphere hepatic brachytherapy.

METHODS AND MATERIALS: Members of the Radioembolization Brachytherapy Oncology Consortium met as an independent group of experts in interventional radiology, radiation oncology, nuclear medicine, medical oncology, and surgical oncology to identify areas of consensus and controversy and to issue clinical guidelines for Y90 microsphere brachytherapy.therapies.

CONCLUSIONS: Yttrium-90 microsphere therapy is a complex procedure that requires multidisciplinary management for safety and success. Practitioners and cooperative groups are encouraged to use these guidelines to formulate their treatment and dose-reporting policies.

- [European Neuroendocrine Tumor Society \(ENETS\) 2007 Consensus Guidelines for the Management pf Patients with Digestive Neuroendocrine tumors](#) (Abstracts)

Part1 Stomach, Duodenum and Pancreas

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Neuroendocrinology 2006 Vol 84 No 3
Publisher Karger: published online, February 2007

- [Neuroendocrine Tumors: A comprehensive Guide to Diagnosis and Management](#) (PDF file - Full text 259 pages)

Gregg Mamikunian, Aaron I. Vinik, MD, PhD, FCP, MACP; Eugene A. Woltering, MD, FACS; Thomas M. O'Dorisio, MD; Vay Liang W. (Bill) Go, MD

Published by [Inter Science Institute 2009](#).

This book adds a new dimension to patient diagnosis, management and monitoring, not only through powerfully discriminating assays, but through the recognition of clinical presentations and syndromes.

To print the full text 259 page PDF file [click here](#), or if you prefer you can order the book or a CD from [Inter Science Institute](#).

For book/CD requests, use this e-mail address: requests@interscienceinstitute.com

- [Guidelines for the management of gastroenteropancreatic neuroendocrine \(including carcinoid\) tumours](#) (PDF Full text)

Ramage JK, Davies AH, Ardill J, Bax N, Caplin M, Grossman A, Hawkins R, McNicol AM, Reed N, Sutton R, Thakker R, Aylwin S, Breen D, Britton K, Buichana K, Corrie P, Gilliams A, Lewington V, McCance D., Meerhan K, Watkinson A:

UKNET work for Neuroendocrine Tumours.
GUT 2005 Jun;54 Suppl 4iv11-6

- [Guidelines for the diagnosis and management of carcinoid tumours, part 1: the gastrointestinal tract](#) (PDF Full text)

A statement from a Canadian National Carcinoid Expert Group

J. Maroun MD,* W. Kocha MD, L. Kvols MD,
G. Bjarnason MD, E. Chen MD, C. Germond MD,
S. Hanna MD, P. Poitras MD, D. Rayson MD,
R. Reid MD, J. Rivera MD, A. Roy MD, A. Shah MD,
L. Sideris MD,|| L. Siu MD, and R. Wong MD

Current Oncology - Volume 13, number 2, 2006

- [Guidelines for the Diagnosis and Treatment of Neuroendocrine Gastrointestinal Tumours](#) (PDF Full Text)

A Consensus Statement on Behalf of the [European Neuroendocrine Tumor Society \(ENETS\)](#)

U. Plöckinger, G. Rindi, R. Arnold, B. Eriksson, E.P. Krenning, W.W. de Herder, A. Goede, M. Caplin, B. Wiedenmann, K. Öberg, J.C. Reubi, O. Nilsson, G. Delle Fave, P. Ruszniewski, H. Ahlman
Neuroendocrinology 2004;80:394-424

Published online: April 18, 2005

[Publisher Karger](#)

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Published on The Carcinoid Cancer Foundation (<http://www.carcinoid.org>)

- [ENETS 2007 Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine tumors. Part1 Stomach, Duodenum and Pancreas](#)

[ENETS Working Group Sessions, November 3, 2005 \(Full Text\)](#)

[Endocrine Tumors of the Duodenum](#)

[Gastric Tumor 1](#)

[Gastric Tumor 2](#)

[Pancreatic Tumors 1](#)

[Pancreatic Tumors 2](#)

[Pancreatic Tumors 3](#)

[Pancreatic Tumors 4](#)

[Poorly Differentiated Endocrine Carcinomas](#)

- Guidelines for the Management of Gastroenteropancreatic Neuroendocrine Tumours (Including Bronchopulmonary and Thymic Neoplasms)

Members of Nordic the NE Tumour Group: Öberg K, Astrup L, Eriksson B, Falkmer UG, Gustafsen J, Haglund C, Knigge U, Vatn M H and Välimäki M.
Acta Oncologica Vol.43, No 7, 2004

Part I-general overview (pp. 617-625) [Full Text](#)

The incidence of neuroendocrine tumours of the gastroenteropancreatic system seems to have increased during the past decade. New diagnostic and therapeutic procedures have aroused the interest of physicians, though most see very few cases of such diseases. A group of members of the Nordic Neuroendocrine Tumour Group decided to compile some guidelines to facilitate the diagnosis and treatment of patients with these tumours. Part I of these guidelines discusses the principles of histopathology, biochemical and radiological diagnosis as well as therapeutic options.

Part II-specific NE tumour types (pp 626-636) [Full Text](#)

Part II of the guidelines contains a description of epidemiology, histopathology, clinical presentation, diagnostic procedure, treatment, and survival for each type of neuroendocrine tumour. We are not only including gastroenteropancreatic tumours but also bronchopulmonary and thymic neuroendocrine tumours. These guidelines essentially cover basic knowledge in the diagnosis and management of the different forms of neuroendocrine tumour. We have, however, tried to give more updated information about the epidemiology and histopathology, which is essential for the clinical management of these tumours.

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- [Consensus Report on the Use of Somatostatin Analogs for the Management of Neuroendocrine Tumors of the Gastroenteropancreatic System](#) (PDF Full text)

By K. Öberg, L. Kvols, M. Caplin, G. Delle Fave, W. de Herder, G. Rindi, P. Ruzsniowski, E. A. Woltering, and B. Wiedenmann
Annals of Oncology 15: 966-973, 2004

This consensus report gives a detailed description of the use of somatostatin analogs in the management of neuroendocrine tumors of the gastroenteropancreatic system. As background information we have outlined critical aspects of the pathology, the use of tumor markers, a definition of functional and non-functional digestive neuroendocrine tumors, different imaging modalities, surgical considerations, liver embolization and the use of cytotoxic drugs as well as interferon. Included in the report is an overview of somatostatin, somatostatin analogs and its receptor expression in different neuroendocrine tumors. It will also define the binding affinities of different somatostatin analogs to the five different subtypes of somatostatin receptor. We compare the

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efficacy of octreotide and lanreotide in reducing diarrhea and flushing. Side-effects are described and we provide practical information on somatostatin analog treatment.

- [Procedure Guideline for Somatostatin Receptor Scintigraphy with In-111 Pentetreotide](#) (PDF)

Version 1.0, approved February 21, 2001, by [Society of Nuclear Medicine](#)

In-111 pentetreotide is a [In-111 DTPA-D-Phe-] conjugate of octreotide, a long-acting somatostatin analog (OctreoScan®).

- [Standardisation of Imaging in Neuroendocrine Tumours: Results of a European Delphi Process](#)

This link is to the PubMed abstract. From the abstract there are direct lines to the full text article on [European Journal of Radiology](#) website (requires subscription payment of membership in a medical society) and the [Science Direct](#) website (requires subscription payment.)

By J. Ricke, K., J. Klose, M. Mignon, K. Öberg, and B. Wiedenmann
European Journal of Radiology, Volume 37, Issue 1, January 2001, Pages 8-17

Diagnostic imaging; Multiple endocrine neoplasia; Neuroendocrine tumours

In 1998 and 1999, a delphi consensus procedure was performed to establish guidelines for standardised diagnostic imaging of neuroendocrine tumours. The procedure included four consecutive workshops of a European group of experts in neuroendocrine tumours as well as feedback given by specialists from the departments of radiology, nuclear medicine, surgery and internal medicine of the according home institutions. Diverging approaches among the centres, which became apparent during the discussion, reflect a lack of controlled studies specifically for rare subgroups of neuroendocrine tumours. This paper summarises the standards for diagnostic imaging as developed during the delphi process. In particular, the diagnostic workflows as well as the technical properties of different imaging modalities are described in detail.

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