

ENETS 2011 Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Tumors: An Update

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Why These Second ENETS Guidelines?

Several guidelines on the management of neuroendocrine tumors have been published by expert national and international groups in recent years [1–6], however, a consensus on diagnosis and treatment is difficult to reach due to the limited evidence available in the literature. Nonetheless, novel diagnostic tools and therapies have emerged in the last 5 years as a substantial result of the continuous effort in the field [7–11]. Indeed, slowly but constantly the neuroendocrine tumor world moves forward for the patients' good.

The need for universal standards has inevitably emerged. This is well exemplified by the journey toward a common tumor grading and staging after the ENETS proposal [7, 8]. Today the International Union Against Cancer (UICC), the American Joint Cancer Committee (AJCC) and the World Health Organization (WHO) substantially endorsed the ENETS proposal [12–14]. A common language is at the basis of this internationally accepted classification. Its simple rules are: (i) the adjective 'neuroendocrine' is defined to specifically connote this neoplastic disease, recognizing the expression of neuroendocrine markers in tumor cells; (ii) the word 'neoplasms' is defined to embrace the whole family of low-,

intermediate- and high-grade tumors; (iii) the term 'tumor' (neuroendocrine tumor, NET) is meant for low- to intermediate-grade neoplasms, as previously defined either 'carcinoid' or 'atypical carcinoid'; (iv) the word 'carcinoma' (neuroendocrine carcinoma, NEC) is meant only for high-grade neoplasms, as previously defined poorly differentiated carcinomas. This terminology is adopted by the ENETS 2011 Guidelines.

The Third Event

In November 3–5, 2010, the European Neuroendocrine Tumor Society (ENETS) held its third Advisory Board meeting in Barcelona aiming at critically discussing and updating the ENETS Guidelines on the Diagnosis and Treatment of Neuroendocrine Tumors generated in 2005–2006 [5, 6].

The consensus sessions covered the following neuroendocrine neoplasm-related topics by sites of origin or stage: gastroduodenal, hindgut, functional pancreatic, non-functional pancreatic, midgut (including appendix), and a final session that covered liver and other distant metastases from neuroendocrine neoplasms of any origin.

How We Worked

Participants at the conference were asked to focus on the relevant literature published between 2006 and 2010. They met over two and a half days, in which data and new evidence were presented. The participants then retreated to break-out sessions according to their disciplines and were required to answer questions listed in a workbook created by the session chairs and the organizing committee. The workbook questions were tailored on the text of the ENETS 2005–2006 Guidelines but focusing on the new available evidence.

The workbooks had essentially three parts: a minimal consensus section from the Guidelines generated in 2005–2006; a reference section with the relevant updated literature as selected by the chairpersons with the aid of a professional librarian bibliographic search, and the questions that the chairpersons considered appropriate to discuss the new evidence. The working groups covered the fields of pathology, imaging, medical and radiotherapy, and surgery. Given the magnitude of the challenge, each working group had two chairpersons with shared obligations. These obligations included: (i) to present the data relevant for updating to the session topic; (ii) to analyze the references generated by the bibliographic search for new potential information, and (iii) to produce the questions and recap the consensus answers to the questions.

All participants were encouraged to challenge the document. Recent data on new evidence and insights were intensely discussed in working group sessions, as well as during the plenary session. Notes were taken continuously so that the final agreement on each question was noted and returned to each session chair for preparation of the consensus statements. The magnitude of the consensus for each answer was estimated. When unanimity was not reached on the final recommendation, the level of consensus was noted into two categories: almost unanimity (90% or more) or majority (more than 50%).

The next step was to review the data produced and to transform it into working papers for publication. The Organizing Committee defined a specific protocol establishing the design of each paper, the tasks for authors, and the general authorship policy. The papers were designed to update the previously published ENETS Guidelines, incorporating the approved consensus statements.

Achievements and Final Remarks

The following six papers are a significant and tangible result of the Consensus Conference. These papers update the work made by ENETS and by all participants at the two former consensus conferences held in Frascati, and adapt to clinical practice the most recent evidence on NET's management.

All participants contributed a great effort equally and delegates generously devoted their time, experience and enthusiasm to building the following consensus guidelines. We thank them for their dedication and good will. We believe that the following papers will be practical and useful instruments for all professionals dealing with patients with digestive NETs. These consensus guidelines underline the possibility of achieving practical standards in such a complex tumor disease and should provide a good framework for patient management and aid in directing future investigative efforts.

Complete List of Participants

List of Participants of the Consensus Conference on the 2011 Consensus Guidelines for the Management of Patients with Digestive Neuroendocrine Tumors: An Update

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