What are Neuro Endocrine tumors (NET)

A NET is a rare type of cancer originating from neuroendocrine cells, which are located all over the body. Therefore the tumor may originate anywhere in the body but most tumors originate in the gastrointestinal tract, the pancreas and lungs. Neuroendocrine cancer can be divided into three groups, based on behavior (the speed of growth). It is thus of utmost importance to hear from your specialist which type of NET or NEC you have. The type of tumor and the speed with which it is growing will determine the kind of treatment possible and the chances of survival.

Research
Through a tissue sample (biopsy) the type of NET can be determined: grade 1 or grade 2. In addition, scans and other research can used to see whether it has spread, if there are metastases, and where all of the secondary tumors are located in the body.

Primary tumor and metastases
The place where the tumor originated is called the place of the “primary tumor”. Small cells may detach from the primary tumor. These cells can spread throughout the body via the blood and lymph vessels. If these cells continue to grow in other places, this is called metastases.

<table>
<thead>
<tr>
<th>Grade 1 NET</th>
<th>Grade 2 NET</th>
<th>Grade 3 NEC</th>
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</thead>
<tbody>
<tr>
<td>Neuroendocrine tumor</td>
<td>Slow growing</td>
<td>Neuroendocrine carcinoma</td>
</tr>
<tr>
<td>Well differentiated</td>
<td>Medium differentiated</td>
<td>Fast rate of growth</td>
</tr>
<tr>
<td>Functional Non functional</td>
<td>Functional Non functional</td>
<td>Poorly differentiated</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Small cells SCNEC Large cells LCNEC</td>
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**Functional and non-functional NET**

NET grade 1 and NET grade 2 can be divided into functional and non-functional based on whether the tumor is producing an excess of hormones or hormone-like substances. (This has nothing to do with sexual hormones). This is more about hormones such as serotonin, neuro-peptides, insulin and gastrin.

*Functional NET*

NET that produce an excess of hormones or hormone-like substances are called ‘functional tumors. If too many hormones or hormone-like substances are produced, then problems may develop. The symptoms can be: hot flushes, anxiety, diarrhea, nausea, dizziness, itching, weight loss, heartburn, heart palpitations and (abdominal) pain. The tumors that cause these symptoms include: NET with carcinoid syndrome, gastrinoma, insulinoma, glucagonoma and vipoma.

*Non functional NET*

If a NET does not produce extra hormones or hormone like substances then it is called a “non-functional” tumor. Non functional NET are often only detected when there is pain. This pain can be caused by the size of the tumor and the place where it is growing. Organs can sometimes be constricted by a growing tumor.

**Treatment options**

Recovery from NET is only possible if all of the tumor is removed during surgery. If recovery is no longer possible, then there are still a number of treatments available. These treatments are aimed at reducing symptoms, prolonging life and improving the quality of life. Which kind of treatment is most appropriate depends on the individual situation. In addition, it is important for patients to consider which treatment they do or do not want to undergo.

The choice of treatment of a NET depends on:

- the patient’s condition
- the growth rate of the tumor and the metastases
- the place where the NET originated (primary tumor)
- the size of the tumor
- the size, location and the number of metastases
- whether the tumor is producing hormones
- the type of hormones that the NET is producing

**Rare tumors**

On average a GP only sees one NET patient during his/her professional career. This is the principle reason why the symptoms are often not immediately recognized by a GP. In addition, NET are sometimes difficult to recognize. This is because the symptoms are often similar to those of more harmless conditions; such as the menopause, asthma or irritable bowel syndrome. Many patients may have symptoms for years before NET is diagnosed.
Specialist treatment centers for NET

Because NET is very rare it is of vital importance to see an experienced NET specialist who has treated many patients with NET. It is also important that your specialist works together in a team with other specialists who have experience with NET patients. These teams are called ‘multidisciplinary teams’. In multidisciplinary teams your situation will be discussed by specialists who are all involved in the treatment. These include an internal medicine specialist, a medical oncologist, an endocrinologist, a pulmonary specialist, a radiologist, a surgeon, a nuclear medicine physician, a radiotherapist and a specialized nurse. Only NET specialist centers have teams with a wealth of experience in the treatment of NET.

Nurse specialists

In hospitals which specialize in NET, nurse specialists are also available. You can easily ask them all of your questions. Nurse specialists can also give you tips on things like pain, shortness of breath, diarrhea, anxiety and insomnia.

Questions for your specialist

It is important to be open in your discussions with the specialist (nurse.) It is quite understandable that it is not always easy to remember all of the information. Therefore make a list of all your questions so you can discuss these during the next visit. A doctor does not mind at all, if you come with a list of questions.

Points to discuss with your specialist:

- Where is the primary tumor?
- Do I have metastases in lymph glands or organs?
- How far the cancer has progressed and what this means in my situation?
- How much experience do you have with the treatment of NET?
- How many patients with NET have you treated?
- Do you have contact with a NET specialist? If so, with whom?
- Are other studies required before the treatment can be started?
- What choices in treatments do I have?
- Which treatment would you recommend, in what order and why?
- What risks or side effects could the proposed treatment have?
- Are there any studies (trials) of new medicines or treatments for NET? In which hospital?

Discuss with the doctor who you can contact, if you have any questions or if there are new problems.

More tips about questions which you may wish to discuss with your specialist can be found on the website of the NET-groep.
Extra help
Once a specialist has told you that you have a NET tumor it can come as quite an emotional shock. Everyone reacts differently to bad news. Some people get sad or angry and want to talk with everyone about it. Others retreat and want to deal with it on their own or share it with their loved ones. There are social workers and psychologists who are specialized in helping cancer patients. If you need this kind of help then your specialist(nurse) or GP can refer you to them.

Think zebra
The zebra is the international symbol for NET. This is because, during their training, doctors learn that when you hear hoof beats this must be a horse. However sometimes this can be from the far less common zebra. In addition to this, each zebra has a unique pattern of stripes. Each and every zebra is different, so is NET!

The NET-groep foundation
The NET-groep advises and supports patients as well as their relatives, children, friends, etc. You are most welcome to come with your questions and worries. We appreciate it when people become members. Enrolment is free via the website or telephone. With more and more members the NET-groep can better represent the interests of patients.

The NET-groep:
- has an informative web-site [www.net-kanker.nl](http://www.net-kanker.nl)
- organizes informative meetings with professionals
- produces information material (films, newsletters and brochures)
- has an open Facebook site with constantly updated news and accessible without a Facebook account
- Supports a private group for patients on Face book (accessed by invitation only)

Board of advisors
The NET-groep has a Board of Advisors of specialists who are of (inter) national importance in the development of treatments for NET. The names of the specialists and the hospitals where they work can be found on the home-page of the NET-groep. Through the knowledge and experience of these specialists the NET group is able to keep you updated on the latest developments around NET.

Financing NET-groep
The NET-groep activities are financed by voluntary contributions from patients and their families and relatives, from funds and (pharmaceutical) companies. The NET group only accepts donations and subsidies when they are given with no intention of profit making. In this way we can avoid a conflict of interests.