Natural History of Familial Carcinoid Tumor

This study will evaluate members in families with a history of small bowel carcinoid cancer to study the natural history of those family members that have the disease, determine ways to improve early detection by performing surveillance on those at risk but without disease and to identify the gene(s) that may cause the tumors. Familial carcinoid tumors usually originate in hormone-producing cells that line the small intestine or other cells of the digestive tract. The tumors are slow-growing and usually take many years before they cause symptoms. It is known that these tumors occur more often in some families and are then passed from one generation to the next by inherited genes.

Members of families, including all siblings and offspring in which two or more immediate blood relatives have had small bowel carcinoid tumors are eligible for this study. In some cases unaffected spouses of family members diagnosed with carcinoid cancer are also requested to participate by donating a sample of blood only.

Participants undergo a medical evaluation every 3 years during a 3- to 5-day hospital stay at the NIH Clinical Center. All participants have a personal and family medical history obtained and undergo a physical examination, blood and urine tests.

People who already have a small bowel carcinoid tumor or are at risk of developing a carcinoid tumor have some or all of the following procedures to determine the presence of carcinoid tumor and its (omit next two words- location or) spread to other areas of the body:
- Video Capsule Endoscopy: Visualization of the gastrointestinal tract by ingesting a disposable, vitamin-pill sized video capsule that has its own camera and light source.
- CT of the chest abdomen and pelvis with oral and IV contrast: X-ray examination of the chest, abdominal and pelvis organs.
- 18 FDOPA Positron emission tomography (PET) with CT for localization: Nuclear imaging scan to look at tumor activity.
- MRI Liver with contrast - to determine if disease has spread to liver
- Gallium 68 PET/CT-limited to individuals that have residual tumor.
- Clinical and research blood work

Should mid gut carcinoid tumors be found every participant will be assisted in determine what the best course of treatment will be for them.

<table>
<thead>
<tr>
<th>Condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carcinoid</td>
</tr>
</tbody>
</table>

Study Type: Observational
Study Design: Time Perspective: Prospective

Official Title: Natural History of Familial Carcinoid Tumor

Resource links provided by NLM:

MedlinePlus related topics: Cancer Carcinoid Tumors

Genetic and Rare Diseases Information Center resources: APUDoma Carcinoid Tumor Neuroepithelioma

U.S. FDA Resources

Further study details as provided by National Institutes of Health Clinical Center (CC):

Primary Outcome Measures:

- The natural history of carcinoid tumor [ Time Frame: On going ] [ Designated as safety issue: No ]

Estimated Enrollment: 99999999

Study Start Date: March 2008
Detailed Description:
Carcinoid tumors are rare and cause either no or few nonspecific symptoms. Therefore, patients with carcinoid tumors most often present late in the course of their illness when there is already progression to an incurable state as a result of metastatic disease. At present there are neither practical population screening tests nor effective therapies and hence the 5 year survival rate is low. Due to the rareness of sporadic carcinoid tumors, large scale genetic analysis and development of sensitive and specific diagnostic tests have not been successful. While kindreds with familial carcinoid tumors that are not ascribable to known genetic syndromes are exceedingly rare, they provide a unique opportunity to facilitate the identification of the responsible gene mutation. In addition, the mutated gene in the rare familial form may also underlie the origin of the more common sporadic occurrence of carcinoid tumors. We propose to study families in which there are at least two known affected members with carcinoid tumors. We aim to diagnose patients with early and therefore potentially curable occult disease. Therefore, family members who have up to a 50% lifetime risk of harboring a carcinoid tumor will undergo an intensive diagnostic evaluation using biochemical, endoscopic and imaging modalities at initial and subsequent two year follow up encounters. Early phenotypic assignment of affected family members and collection of germline and tumoral DNA from multiple kindreds should also facilitate the genetic analysis leading to the identity of the disease gene. Evaluation of affected family members at varying stages of disease will contribute to our understanding of the natural history of carcinoid tumors and the relative utility of a variety of diagnostic and surveillance tests. Hopefully, such knowledge gained will also be applicable to patients with carcinoid tumors occurring sporadically or in the setting of other familial cancer syndromes. There is no planned treatment for patients with existing or newly diagnosed primary or metastatic carcinoid tumors. However, these patients may be evaluated by consultation with oncology and surgery for potential treatment on their service under their preexisting protocols.

Eligibility

Ages Eligible for Study: 18 Years and older
Genders Eligible for Study: Both
Accepts Healthy Volunteers: No

Criteria

- INCLUSION CRITERIA:
  a. Adult patients who are members of families in which there are at least two immediate relatives from consecutive generations that have been diagnosed with gastrointestinal carcinoid tumors (affected family member) and either have a carcinoid tumor or are at a 50% lifetime risk of developing a carcinoid tumor by virtue of being a first degree relative of an affected family member. This includes family members of patients with carcinoid tumors even if the patients with tumors are unwilling to participate so long as we have appropriate documentation confirming the diagnosis of the carcinoid tumors in the affected family members. Adult patients who are unable to provide informed consent but whose wishes suggest they are willing to donate samples for research purposes will be considered for study enrollment.
  b. Unaffected spouses of patients with a carcinoid tumor and who have children.

EXCLUSION CRITERIA:

1. Families with multiple endocrine neoplasia (MEN) I, MEN II or other familial tumor syndromes such as Von Hippel Lindau Syndrome and
Neurofibromatosis for which there is a known genetic predisposition to non-carcinoid tumors as well as carcinoid tumors will be excluded from the study.

2. Pregnancy, breastfeeding.

3. Anticoagulation, seizures, severe systemic disease of any sort, advanced metastatic carcinoid may be relative exclusion criteria prohibiting a full evaluation as described above under protocol design. However, these medical conditions should not absolutely exclude participation in the protocol. Participation in each protocol delineated evaluation procedure will be judged on a case by case basis with patient safety as the paramount consideration.

Contacts and Locations

Choosing to participate in a study is an important personal decision. Talk with your doctor and family members or friends about deciding to join a study. To learn more about this study, you or your doctor may contact the study research staff using the Contacts provided below. For general information, see Learn About Clinical Studies.

Please refer to this study by its ClinicalTrials.gov identifier: NCT00646022

Contacts

Contact: Stephen A Wank, M.D.  (301) 496-4202  stevew@bdg10.niddk.nih.gov

Locations

United States, Maryland

National Institutes of Health Clinical Center, 9000 Rockville Pike
Bethesda, Maryland, United States, 20892
Contact: For more information at the NIH Clinical Center contact Patient Recruitment and Public Liaison Office (PRPL)  800-411-1222 ext TTY8664111010  prpl@mail.cc.nih.gov

Sponsors and Collaborators

National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK)

Investigators

Principal Investigator:  Stephen A Wank, M.D.  National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK)

More Information
Additional Information:

NIH Clinical Center Detailed Web Page

Publications:


ClinicalTrials.gov Identifier: NCT00646022
Other Study ID Numbers: 080098, 08-DK-0098
Study First Received: March 26, 2008
Last Updated: December 2, 2014
Health Authority: United States: Federal Government

Keywords provided by National Institutes of Health Clinical Center (CC):
Neuroendocrine
PET
Gastrointestinal
Serotonin
Carcinoid Tumor
Gastrointestinal Carcinoid Tumor
Familial Cancer Tumor

Additional relevant MeSH terms:
Apudoma
Carcinoid Tumor
Neoplasms by Histologic Type
Neoplasms, Germ Cell and Embryonal
Neoplasms, Glandular and Epithelial
Neoplasms, Nerve Tissue
Neuroectodermal Tumors

ClinicalTrials.gov processed this record on April 15, 2015