National Familial Pancreas Tumor Registry
http://www.pathology.jhu.edu/pancreas

NFPTR NEWS

Volume 1 Issue 1 December, 2000

Visit our site:

www.pathology.jhu.edu/pancreas

Our website includes:

- Active chat room for the patients and families
- Latest pancreas cancer research
- Illustrated pancreas cancer FAQ
- Discussions with social worker
- Links to other sites

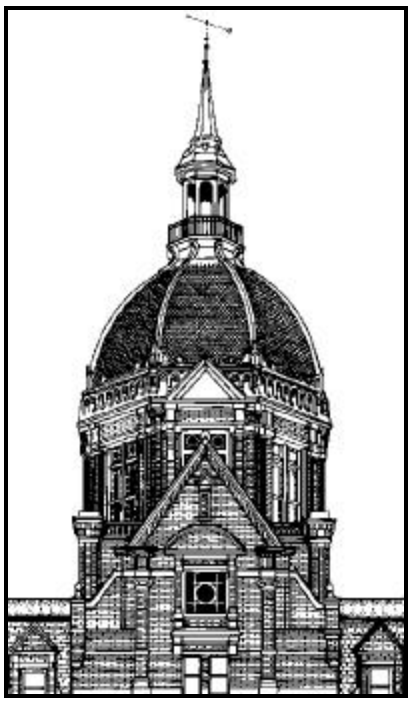
Inside this issue:

UPDATE FROM THE DIRECTOR

Many of you have asked to be periodically updated on the activities of the National Familial Pancreas Tumor Registry (NFPTR). We have therefore created this newsletter, NFPTR News. While the primary goal of NFPTR News is for us to let you know what we've been doing in the past year, we also welcome contributions from participants in the NFPTR. If you would like to submit something for the next issue, just send us a note or drop us an e-mail.

This has been another remarkable year for the NFPTR and for pancreatic cancer research at Johns Hopkins. Thanks to your help and support, the NFPTR has grown to be the largest registry of familial pancreatic cancer in the world. The registry now includes over 500 families! These families provide a critical resource for our studies of why pancreatic cancer runs in some families.

To date, five distinct syndromes have been identified which predispose individuals to pancreatic cancer and for which the genetic basis has been discovered. For those of you who would like to learn more about these five syndromes, we've summarized them on page 4.



The vast majority of patients do not fit into one of these rare clinical syndromes, therefore, the work of the NFPTR goes on!

Continued on bottom of page 2

KEEP IN TOUCH!

When one or two members in a family have been diagnosed with pancreatic cancer, the other members of the family frequently come to us and ask for an assessment of their personal cancer risk. We get questions, such as "my mother and brother have been diagnosed with pancreatic cancer, am I at an increased risk of getting pancreatic cancer?" As discussed on pages 2 & 3, the NFPTR is currently conducting several studies to see if other individuals in families where there has been a pancreatic cancer are themselves at an increased risk for developing the disease. In order to help complete these studies, it would be greatly appreciated if you would complete the enclosed form and return it to us in the enclosed self-addressed stamped envelope.

These studies will form an important basis for predicting cancer risk and for developing new tests to screen for early detection.

SCREENING FOR PANCREATIC CANCER USING ENDOSCOPIC ULTRASONOGRAPHY

Dr. Mimi Canto at Johns Hopkins has recently been awarded a grant to study the usefulness of endoscopic ultrasonography (EUS) in the early detection of pancreatic cancer. As a part of this research study, Dr. Canto is currently screening selected healthy family members from the NFPTP. (primarily individuals in families where three or more pancreatic cancers have been diagnosed) using EUS.

EUS is a technique in which a small endoscope is inserted into a patient's mouth, passed down through their stomach and into the small intestine. When placed directly into the small intestine, the ultrasound transducer at the tip of the scope can be used to provide detailed images of the pancreas. Dr. Canto is currently studying the accuracy of this approach in detecting small, and therefore potentially curable, pancreatic cancers.

While this is a pilot research project, we are excited because this is one of the first studies to test the effectiveness of this relatively noninvasive procedure in the early detection of pancreatic cancer.

Mimi Canto, M.D. M.H.S.

If you would like to learn more about Dr. Canto's study, please e-mail her at mimicanto@aol.com

NEW NFPTR COORDINATOR: KIERAN BRUNE

As some of you may know, Florence Falatko has stepped down as Coordinator for the NFPTR, so that she can spend more time with her family. Kieran Brune joined us as the new Coordinator of the NFPTR in June 2000. After graduating from Boston College with a Bachelors of Science in Biology, she worked for two years as a research technician in the Neurology Department at Johns Hopkins.

Kieran's daily contact with families who have faced pancreatic cancer makes her a very dedicated team member in the search for a cure for this dreaded disease.

Please feel free to call her at 410-955-3502 or email her at kbrune@jhmi.edu with any questions you may have or just to say hello. Should any new cancers develop in the family, please let Ms. Brune know.

Kieran Brune, Coordinator

…UPDATE (continued)

In addition to hunting for the genes which are responsible for familial pancreatic cancer, we are also currently conducting several studies to see if apparently healthy individuals from families in which there has been a pancreatic cancer are themselves at an increased risk for developing this disease.

Therefore, we would greatly appreciate it if you would take time to complete the enclosed form which asks for an update of your family's health status.

Your cooperation in this endeavor, as always, is greatly appreciated.

We hope you enjoy this newsletter and we welcome your feedback. Our sincerest wishes for Happy Holidays and a healthy New Year from the staff of the NFPTR!

Ralph H. Hruban, M.D., Director NFPTR

"Nothing in life is to be feared, it is only to be understood."

Marie Curie

LABORATORY FOR THE EARLY DETECTION OF PANCREATIC CANCER

With the generous support of the National Pancreatic Cancer Advocacy Group (PanCAN) and the Michael Rolfe Foundation for Pancreatic Cancer Research, Dr. Michael Goggins opened an exciting new laboratory at Johns Hopkins dedicated to developing an early detection test for pancreatic cancer.

Dr. Goggins' approach is simple. He believes that by understanding fundamental genetic and protein changes that are found in pancreatic cancer he will be able to identify a specific marker for the early detection of pancreatic cancer.

Dr. Goggins' lab has already discovered a number of genetic alterations in pancreatic cancer that may be markers for the disease. These alterations are called "hypermethylation" and Dr. Goggins' laboratory is now working hard to further characterize these alterations (see Cancer Research volume 60, number 7, pages 18351 839, 2000).

In addition to these genetic changes, Dr. Christophe Rosty in Dr. Goggins' lab is using a new "protein chip" technology to identify proteins which are secreted by pancreatic cancers.

Just as there is a PSA test for prostate cancer, so too is Dr. Goggins' laboratory dedicated to finding an effective screening test for the early detection of pancreatic cancer.

Christophe Rosty, M.D. and Michael Goggins, M.D. in the Early Detection Lab

JENNIFER SOLLENBERGER: NEW GENETIC COUNSELOR

We are pleased to announce the addition of Jennifer Sollenberger to the NFPTR Team. Jennifer joined us as the genetic counselor for the registry in June of 2000. Ms. Sollenberger has a Masters of Science in Genetic Counseling from the University of Colorado as well as a Masters of Science in Molecular Biology and Genetics from Northwestern. Ms. Sollenberger specializes in cancer genetics. As part of her work with the NFPTR, she will provide a genetic counseling session to those individuals who participate in Dr. Canto's screening study.

If you would like to learn more about genetic counseling for your family, you can reach her by phone at 410-614-0378 or e-mail at solleje@jhmi.edu.

Jennifer Sollenberger, M.S.

NEW STUDY ON HOW PANCREATIC CANCER IS INHERITED

Alison Klein, a Ph.D. student here at Johns Hopkins University, is studying the families enrolled in the NFPTR as part of her doctoral thesis. She is applying computer models to the family information we have collected in the registry to gain a better understanding of how pancreatic cancer is inherited in some families. Through Ms. Klein's work, we will learn what percentage of the population may have a yet to be discovered "pancreatic cancer gene" and how many people who have this gene go on to develop this disease.

The results of this important study will help us to develop future studies to find the genes that increase the risk of pancreatic cancer so that we may be able to improve the counseling, screening, and treatment for pancreatic cancer.

Alison Klein, M.H.S.

NATIONAL FAMILIAL PANCREAS TUMOR REGISTRY
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BEST WISHES FOR A HAPPY HOLIDAY SEASON FROM THE STAFF OF THE NFPTR!

WHAT EVERYONE SHOULD KNOW ABOUT PANCREATIC CANCER

- There is always reason for hope!
- Pancreas cancer is the fourth leading cause of cancer death for both men and women.
- Pancreas cancer is difficult to diagnose because of the location of the organ.
- Pancreas cancer is treatable when detected early.
- Five year survival rate approaches 40% if cancer is caught before it spreads and is surgically removed.
- Survival rate after surgery for pancreatic cancer is higher at specialized hospitals, such as Johns Hopkins, than it is at other hospitals that rarely do pancreas surgery.
- Symptoms of pancreas cancer include back and abdominal pain as well as yellowing of the skin (jaundice).
- Smoking increases your risk for pancreas cancer as well as other types of cancer.

GENETIC SYNDROMES

Five distinct syndromes have been identified which predispose individuals to pancreatic cancer and for which the genetic basis has been discovered. These syndromes are:

1) Familial Breast Cancer
Inherited mutations in the second breast cancer gene, called "BRCA2," have been shown to predispose family members to both breast and pancreatic cancer. This syndrome is rare, but is more common in individuals of the Ashkenazi Jewish descent, in which it strikes approximately 1% of this population.

2) Familial Atypical Multiple Mole Melanoma Syndrome (FAMMM)
FAMMM is a very rare syndrome caused by inherited mutations in the p16 gene. Patients with this disorder not only have many skin moles, but they aiso have an increased risk of developing a form of skin cancer called melanoma and of developing pancreatic cancer.

3) Peutz-Jeghers Syndrome
The Peutz-Jeghers syndrome is a rare syndrome characterized by pigmented spots on the lips and inside of the mouth and by a specific type of intestinal polyp. Patients with the Peutz-Jeghers syndrome have an increased risk (as high as 30% lifetime risk) of also developing cancer of the pancreas.

4) Familial Chronic Pancreatitis
Familial pancreatitis is a rare syndrome characterized by an inherited predisposition to developing inflammation of the pancreas at a young age. Many families with this syndrome have been found to inherit mutations in the "cationic trypsinogen gene".

5) Hereditary Non-Polyposis Colorectal Cancer (HNPCC)
Patients with this syndrome have an increased risk of developing colon cancer at a young age, and some may also have a predisposition to develop pancreatic cancer.

While these syndromes account for only a small percentage of the familial clustering of pancreatic cancer, they are important because genetic testing may be indicated in selected families. If you would like to learn about genetic counseling, please contact Jennifer Sollenberger (410-6140378 or solleje@jhmi.edu) here at Hopkins. If you would like the number of a genetic counselor in your area, you can find the name of a trained counselor by contacting the American Society of Genetic Counselors (telephone: 610-872-7608, or visit their Web site www.nsgc.org/resource_link.html).