NFPTR national familial pancreas tumor registry

NEWS

DECEMBER 2011

FROM THE DIRECTOR

We have had another busy year working hard to make progress against this dreadful disease. Our work would not be possible without the continued support of the families enrolled in the registry. Thank you for your support and your continued involvement in our research.

In our efforts to provide outstanding clinical care for pancreatic cancer patients and individuals at risk of developing pancreatic cancer, we continue to expand our multidisciplinary clinics. In this newsletter we highlight our ongoing Pancreas Multi-disciplinary Clinic now in its fifth year and how this approach has recently expanded to include evaluation of individuals with pancreatic cysts. For more details see our story on page 3.

While improving treatment for pancreatic cancer is critical so is the development of ways to detect pancreatic cancer early, before it becomes invasive. Over the past decades, researchers around the globe have sought to develop early detection methods for pancreatic cancer. This spring, the first international summit aimed at evaluating early detection methods for pancreatic cancer met in Baltimore. At this summit researchers discussed their experiences screening for pancreatic cancer and began discussions for planning future clinical trials for pancreatic cancer screening (page 2).

Understanding how pancreatic cancers develop is an important component to the development of better treatments and can also help to identify individuals at highest risk who may benefit from early detection screening programs, such as those discussed at the recent summit. To that end, we are happy to report that our group received two new grants aimed at identifying the genetics underlying the development of pancreatic cancer. In the first, Dr. Eshleman is using next-generation sequencing approaches to study pancreatic cancer cells from patients with familial pancreatic cancer. (page 3) The second study, lead by Dr. Klein brings together an international team of researchers to identify genetic variation that may increase risk of pancreatic cancer (page 2).

It is our hope that all of these ongoing projects will lead to improved care for patients with pancreatic cancer and management of individuals at high-risk of developing pancreatic cancer.

Best wishes for you and your family in this New Year! We hope that we will continue to make progress against this disease in the year to come!

-Dr. Alison Klein

NFPTR TEAM (left to right): Diane Echavarria (Coordinator), Dr. Alison Klein (NFPTR Director), Irina Usach (Coordinator)

FROM THE COORDINATORS

This year we are 4,200 families-strong and continue to welcome all families affected by pancreatic cancer to join the registry.

We sincerely thank every person that has reconnected with family members, dug into family records, jogged their memories in search of details, and has emailed or called with requests or updates. Your efforts allow us to study family trends and to seek the elusive gene mutations that may be associated with pancreatic cancer. It's because of your kindness and willingness to share your story that we are inspired and continue our research.

We're happy to hear from you and add new family members to our list for next year's newsletter mailing. Thank you for your courage and dedication–and as always, feel free to contact us directly!

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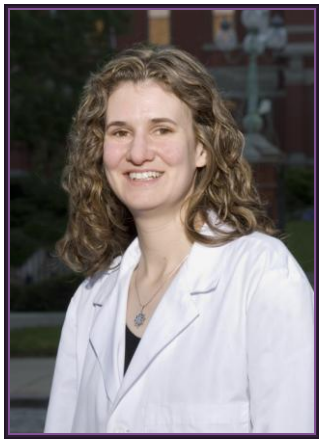
INTERNATIONAL SUMMIT FOR PANCREATIC CANCER SCREENING

An International summit hosted by Johns Hopkins on Cancer of the Pancreas Screening met in Baltimore in the spring of 2011. This meeting brought together leaders in the field of pancreatic cancer screening from Europe, Canada, Asia, and the United States. Dr. Marcia Canto, principal investigator of our Cancer of the Pancreas Screening studies (CAPS), led this meeting. Participants included experts in the fields of medical oncology, gastroenterology, surgery, pathology, and genetics.

Discussions focused on each group's experiences in early detection screening for pancreatic cancer including comparison of screening technologies (i.e. endoscopic ultrasound, MRI, CT-SCAN), how to identify high-risk populations who may benefit from screening and when to operate on individuals who have abnormal screening results. The goal of these discussions is to plan for future clinical trials aimed to determine the effectiveness of early detection screening for pancreatic cancer.

NEW NCI GRANT FOR GENOME-WIDE ASSOCIATION STUDY OF PANCREATIC CANCER

The genetic basis of pancreatic cancer is complex. We know that both genetic and environmental factors such as cigarette smoking can play important roles. Much of our work in the NFPTR is to identify the genetic factors that play a role in pancreatic cancer risk. Even though the role of genetic factors is quite complex with only a portion of the genes involved in pancreatic cancer risk having been identified to date, we are taking several approaches to identify these genes. Last year we announced our Familial Pancreatic Cancer Sequencing consortium, which is using next-generation sequencing technologies to identify familial pancreatic cancer genes in high-risk families.



In a complementary approach this year in a new grant funded by the National Cancer Institute, Dr. Klein is leading an international multicenter team of investigators to conduct a genome-wide association study of pancreatic cancer. Bringing together investigators from the United States, Canada, Europe and Australia, this study will examine genetic variation throughout the genome to identify genetic variants that may be more common in individuals with pancreatic cancer. Through identifying these variants, we may be able to better understand how pancreatic cancer develops as well as identify individuals at highest risk who may benefit from early detection screening programs.

IN THE NFPTR SPOTLIGHT: JAMES R. ESHLEMAN, M.D., PH.D.

Dr. James R. Eshleman's lab focused research this year on analyzing the world's largest collection of familial pancreatic cancer cell lines.

For the past 8 years, Dr. Eshleman's lab has struggled to isolate cell lines from any pancreatic cancer patient with a strong family history. Over the years, his lab has isolated more than 10 of them. These are an invaluable resource to identify genes that cause familial pancreatic cancer, and to find drugs to which they are uniquely sensitive.

Dr. Eshleman's lab is now sequencing 9 familial pancreatic cancers with the hopes of identifying pancreatic cancer susceptibility genes. Any new genes discovered will be tested using a panel of familial patient DNA from the NFPTR to determine frequency. These tumors will also be tested for chemosensitivity against a panel of 3,300 drugs. Not only has the addition of the new fully-sequenced familial pancreatic cancers made our collection the largest in the world, but it's this "dual-pronged approach," searching for mutations and linking chemosensitivity profiles to mutation profiles that Dr. Eshleman hopes will allow us to not only find new mutations but allow us to learn about how these mutations translate to effective treatments for pancreatic cancer.

TWO COMPREHENSIVE CLINICS, ONE VERY DEDICATED CLINICAL TEAM

Previously we reported very exciting details of the now 5 years running Johns Hopkins Pancreas Multidisciplinary Cancer Clinic (MDC) where patients with known non metastatic or suspected pancreatic cancer benefit from consultation with a comprehensive team of renowned providers. Medical and radiation oncologists, surgical oncologists, pathologists, dieticians, diagnostic radiologists, gastroenterologists and geneticists collectively review all imaging, medical history and pathology ultimately agreeing on treatment options for each patient.

Now patients with suspected pancreatic cysts have the opportunity to participate in a similar clinical service!

According to the Multidisciplinary Pancreatic Cyst Program Clinical Coordinator Lindsey Manos, MPAS, PA-C, "Based on the current studies, 8-20% of patients with pancreatic cancer that undergo surgical resection are due to malignant transformation of a cyst. With diligent follow up this is a great opportunity for us to intervene early."

As we learn more about pancreatic cancer and its sometimes cystic precursors and as imaging improves to find both benign and precursor cysts earlier, we're left with important questions: what are the treatment options for pancreatic cysts? Should a cyst be removed or just monitored?

Expert clinicians in the Multidisciplinary Pancreatic Cyst Program, part of the Division of Gastroenterology and The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins hold a weekly clinic to help patients with suspected cysts find answers. Just as the Pancreas MDC provides an "all-inclusive" opinion for care in a 1-2 day clinic the cyst clinic aims to hone this multidisciplinary approach coordinating the necessary tests and analyses of cysts to spell out expert and comprehensive advice for treatment.

For more information on the clinics please visit their Web sites:

http://pathology.jhu.edu/pancreas/MDC/index.php
http://pathology.jhu.edu/pancreas/cyst/index.php

HOW YOU CAN HELP:

Spouses are eligible to donate a saliva sample as a "control" (a person without pancreatic cancer to serve as a comparison) for our research studies. Contact us at pancreas@jhmi.edu with "Control" in the subject line.

Family members with at least one first-degree relative with pancreatic cancer (sibling, parent, or child) as well as another family member with pancreatic cancer on the same side of the family may also be eligible to donate a blood sample. Contact us at pancreas@jhmi.edu with "Blood sample" in the subject line.

Interested in Screening? Individuals with two or more family members with pancreas cancer may be eligible for a research screening study (CAPS4) using endoscopic ultrasound here at Hopkins. For information, please contact the study coordinators, Hilary Cosby or Verna Scheeler at **caps4@jhmi.edu** or 410-502-9795.

PLEASE REMEMBER TO RETURN YOUR UPDATE CARD ENCLOSED WITH THIS NEWSLETTER.
Even if there have been no changes in your family, this information is very important to our research. Thank you!

CERTIFICATE OF CONFIDENTIALITY

We want to remind the participants that the NFPTR continues to be protected by a Certificate of Confidentiality (NCI-01-062) from the National Institutes of Health, Department of Health and Human Services.
This certificate further helps us protect the confidential information that you have provided by giving us legal protection from having to involuntarily release any information about you. With this certificate, we cannot be forced by court order to disclose any information for criminal, administrative, legislative, or other proceedings.

If you have any questions regarding this or would like a copy, please contact Diane Echavarria: (410) 955-3502

MEDICAL DONATION RESEARCH PROGRAM

Dr. Iacobuzio-Donahue's Gastrointestinal Cancer Rapid Medical Donation Program (GICRMDP) continues to gather crucial information about metastatic gastrointestinal cancer by participants who volunteer prior to their death to undergo a rapid, research autopsy. If this research study is something you or a family member would like to learn more about, feel free to contact Dr. Iacobuzio-Donahue at ciacobu@jhmi.edu or call her at (410) 955-3511.

LEARN MORE ABOUT OUR RESEARCH!

Below is a short bibliography of our most recently published research conducted by investigators working with the NFPTR.

You can view abstracts of most or all of these articles by visiting www.pubmed.com and copying and pasting the title of the article into the search field. If you have any questions about any of the studies discussed in this newsletter or listed here, please contact the NFPTR at 410-955-3502 or pancreas@jhmi.edu.

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