Prevention *matters*

| A FOX CHASE CANCER CENTER **RISK ASSESSMENT PROGRAM** PUBLICATION | **SPRING/SUMMER 2015** |

INTERVIEW WITH DON MEADOWS: A Birt-Hogg-Dubé syndrome patient

LISA BEALIN, BS

pictured: Don Meadows and son

Birt-Hogg-Dubé syndrome (also known as BHD) is a hereditary condition named after the three Canadian doctors who first described it in 1977 – Arthur R. Birt, Georgina R. Hogg, and William J. Dubé. BHD syndrome is caused by mutations in the Folliculin gene (FLCN), and only a genetic test can find mutations. BHD can cause skin lesions, lung cysts, collapsed lungs and kidney cancer.

Tell me about the personal and family history that led you to consider genetic testing.

About 18-20 years ago my dad was hospitalized with a heart condition. He was experiencing some indigestion and while having some tests it was found that his kidneys had multiple tumors. His doctors assumed they were cancerous and removed one kidney and part of his other kidney. He eventually needed dialysis. Birt-Hogg-Dubé syndrome (BHD) was not discovered yet, but the doctors thought there could be a genetic link because it was not common to have tumors in both kidneys. As I got older, I visited a urologist and explained my dad's history. We decided to do an ultrasound and then a CT (computed tomography) scan of my kidneys. My left kidney showed a 4 cm tumor. The doctor suggested one more test, an MRI (Magnetic Resonance Imaging), which showed 2 tumors in my left kidney and 3 in my right kidney. My doctor heard Dr. Robert Uzzo speak at a gathering and suggested I see him at Fox Chase. I met with Dr. Uzzo and he suggested that after my surgery to remove my tumors, I should meet with the Risk Assessment program's genetics team.

How were you found to have a Birt-Hogg-Dubé syndrome mutation? Have any other relatives been tested?

I met with genetic counselor, Andrea Forman. We went over my family history and she explained that the benign kidney tumors could be suggestive of BHD. I went ahead with the testing and was found to be positive for the mutation. I was not shocked to hear the results. I am a person who likes to research and get all of the information that I can. None of my relatives have been tested yet. I shared with my son and daughter that they have a 50 percent chance of having the mutation. When my son turns 18 he wants to be tested. I explained to them it is their choice to be tested. My sisters assume they are positive. They have the benign skin lesions (a symptom of BHD). One of my sisters gets kidney scans.

How has having BHD changed your screening and medical care?

I get an ultrasound every 6 months to check my kidneys. I still have small tumors in my kidney which are monitored. Dr. Uzzo explained that there is no need to remove them at this time.

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FOX CHASE CANCER CENTER
TEMPLE HEALTH

HELP RAP GO GREEN

Help RAP go green by receiving **Prevention Matters** electronically. To receive an electronic version of this newsletter rather than a print copy, e-mail us at: **rapinfo@fccc.edu.** Please include your name in the e-mail. ***Thank you.***

IN THIS ISSUE

THE IMPORTANCE OF SHARING

MICHELLE SAVAGE, MS, LCGC

If you had genetic testing, your healthcare team may have suggested you share your test results with family members. A person who tests positive for a genetic mutation may share their results with their closest family members, such as their children and sisters and brothers, but sometimes do not think to share these results with more distant relatives, such as cousins. It may seem like your genetic results would be of little benefit to your distant family members, but this information is actually very important to share. If you have been found to carry a genetic mutation that increases your risk for cancer, each of your first degree relatives has a 50% (1 in 2) chance of also carrying this mutation. First-degree relatives include your children, your brothers and sisters, and both of your parents. We encourage reaching out to your aunts, uncles and cousins as these relatives could also have inherited the same genetic mutation. We may even recommend testing for more distant family members.

Not sharing genetic test results with relatives can impact their ability to get the right medical care. Costly genetic tests might be ordered that are not necessary or they may have surgeries or procedures that are not necessary. In the worst case scenario, a family member may even develop cancer before they find out that they carry the same cancer risk mutation found in the family.

Although sharing these results may seem difficult or uncomfortable, it is important to remember that knowledge is power when it comes to genetic information. By sharing your test results with family members, you may help these relatives reduce their risks for future cancers or find a cancer at an earlier and more treatable stage.

Here in the Risk Assessment Program, we have tools to help our patients share this information with family members. We give our patients brochures with our contact information and reasons why relatives should consider genetic testing. We also provide personal family letters, copies of your genetic test results, as well as copies of the family tree to help share this information with your family. If family members are not local, we can help them find genetic counselors close to their home.

If you would like help sharing your genetic test results with your family members, please call one of our genetic counselors at 877-627-9684. We will be happy to give you some of these materials and discuss ways to share this information with your family.

Continued from front cover

I decided to keep my body healthy and strong. About 3-4 years ago, I changed my eating and began to exercise regularly. I feel that I will be able to recover from any additional surgery if my body is healthy.

What has been the most difficult thing about having BHD?
I feel that I can deal with myself, but the knowledge that this could affect my kids and other family members is difficult.

What resources have been helpful to you?
I like to research and gather information. There is a website www.bhdsyndrome.org that is very helpful. The information is easy to understand.

How has speaking and writing about your experience been helpful? Have you shared your story?
My son and I created a video to help process this together. He is hoping to be a filmmaker. The video will be part of his application for school. We have shared this video with others and it allows people to experience my story. I am a pastor at my church and I have told my story. It has deepened my faith.

Watch the video "Inheritance"
www.vimeo.com/115376125

Is there a message you would like to share?
Difficult experiences can help you connect with the people you love. You don't choose these things, but there is value when you deepen your relationships.

IN THE NEWS

DR. MARY DALY RECEIVES SUSAN G. KOMEN LIGHTS OF LIFE AWARD

Fox Chase Cancer Center and their Breast Cancer Patient Care Team were recently honored at the Komen Foundation's Pink Thai Ball in Philadelphia. Fox Chase was awarded the 2014 Beacon of Hope, and the Fox Chase Breast Cancer Patient Care Team, including Dr. Mary Daly, chair of the Department of Clinical Genetics, received the 2014 Lights of Life award. The awards recognize Fox Chase's pioneering history in cancer care, and the continued leadership and dedication of their faculty and staff.

TOGETHER FACING GYNECOLOGIC CANCER EVENT

The Fox Chase Women's Cancer Center hosted a special event "Together Facing Gynecologic Cancer" on November 15, 2014. Pat Ciarrocchi, CBS 3 reporter and community activist, was the special guest. This event brought together gynecologic cancer survivors, family members, those with a family history of cancer, health professionals, friends and caregivers.

The guests attended various lectures, met the Fox Chase gynecologic oncology team, participated in yoga, Reiki and acupuncture sessions and visited exhibitors' tables. Staff from the Department of Clinical Genetics including genetic counselors Andrea Forman, Kim Rainey and nurse practitioner, Agnes Masny were available to answer questions about the Risk Assessment Program, clinical services and genetic testing. Dr. Mary Daly, chair of the Department of Clinical Genetics, gave a lecture about hereditary cancer risk assessment, and Sharon Schwartz, nurse practitioner, spoke about survivorship issues. About 300 people attended this successful event.

SHARON SCHWARTZ, MSN, CRNP JOINS THE DEPARTMENT OF CLINICAL GENETICS

Sharon Schwartz has joined our department as a nurse practitioner. She works with genetic counselors and patients to review genetic test results and recommendations, and she sees patients in our high risk clinic. She continues to see gynecologic oncology patients. In clinic, she will continue to manage patients' abnormal pap smears. She will also take care of patients with gynecological side effects from their cancer treatment, and see survivorship patients for their follow-up gynecologic care. She is also the oncofertility coordinator for Fox Chase Cancer Center. Sharon earned her undergraduate and graduate degrees in nursing from the University of Pennsylvania, and is certified as a Women's Health Care Nurse Practitioner. She is a board member of the Society of Gynecologic Nurse Oncologists (SGNO). The Risk Assessment Program welcomes Sharon to our staff.

(left to right): Deb Zakrzewski, Loretta Bagden, and Lorraine Crozier

MEET OUR INTAKE TEAM

The Risk Assessment intake team will be happy to help with the following:

- Schedule cancer genetic risk counseling to learn about your risk and if testing is appropriate for you
- Arrange an appointment with a genetic counselor for review of previous testing and/or newly available testing
- Schedule a clinic appointment for women at high risk due to family history of cancer, biopsy finding, or review of genetic test results from outside Fox Chase
- Help you learn if genetic counseling services are part of your insurance plan
- Provide referral information for risk assessment
- Refer you to other services at Fox Chase

Please contact our team at **877-627-9684** or **rapinfo@fccc.edu**.

DIET, INFLAMMATION, AND RISK OF COLORECTAL CANCER IN WOMEN

Inflammation is the immune response of body tissues to injury. It is a normal process that protects and heals the body following physical injury. Inflammation can also become chronic, and this type of long-term process adds to serious illness, such as colorectal cancer. There is growing data that certain foods affect both inflammation and colorectal cancer. Some foods contribute to unhealthy inflammation, while others fight harmful inflammation.

The recent Iowa Women's Health Study looked at the association between diet and colorectal cancer in a large group of 34,703 postmenopausal women ages 55-69 years who were cancer-free at the beginning of the study in 1986.

Researchers from the University of South Carolina built a special Dietary Inflammatory Index (DII) to measure inflammatory potential of diet. A higher DII score reveals a pro-inflammatory diet. Diet that promotes inflammation is one that has its macronutrients out of balance. These are the basic proteins, carbohydrates, and fats that make up the food we eat. Eating too much fried or processed foods, saturated, omega-6 and trans fats, and refined sugar have all been linked to increased inflammation. Women completed a 121-item food questionnaire asking what kind of food they eat and supplements they take. At the end of 2010, researchers found 1,636 cases of colorectal cancer in this group through the State Health Registry of Iowa.

The results of this study showed that women who ate a more pro-inflammatory diet were at higher risk of colorectal cancer compared with women who ate a more anti-inflammatory diet. These results add to the data for the benefits of a diet high in vegetables and fruits, nuts, low-fat dairy, fish, and whole grain, and low in fried foods, deli meats, refined grains, such as white bread, and alcohol. Read more about the study here: http://www.ncbi.nlm.nih.gov/pubmed/25155761

HEALTHY RECIPES

Tomato Soup

- 1 Tbsp. extra virgin olive oil
- 1 cup chopped onion
- 1/3 cup chopped scallions
- 1 (28-oz.) can no-salt added whole tomatoes in tomato sauce
- 3 marinated sun-dried tomato halves, rinsed and chopped
- 1 tsp. dried basil
- 1/2 tsp. sugar
- 1 cup low-sodium tomato juice
- Salt and freshly ground pepper
- Garlic croutons

Heat oil and add onions and scallions. Cook 5 minutes. Add tomatoes one at a time. Add tomato sauce remaining in can. Add sun-dried tomatoes, basil, and sugar. Bring to a boil, reduce heat, cover and simmer for 20 minutes. Purée soup. Blend in tomato juice. Season soup with salt and pepper. Garnish with croutons.

Makes 6 servings.

Per serving: 71 calories, 2.5 g fat (< 1 g sat fat), 12 g carbohydrates, 2 g protein, 2 g fiber, 64 mg sodium.

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Herbed Chickpea Wraps with Grapes and Walnuts

- 1 can (15 oz.) chickpeas (garbanzo beans), drained and rinsed
- 2 cups seedless red grapes, halved
- 1/2 cup finely chopped red onion
- 1/2 cup coarsely chopped walnuts (roasted)
- 1 Tbsp. finely chopped fresh basil
- 1 Tbsp. finely chopped fresh rosemary
- 1 Tbsp. finely chopped fresh thyme
- 1/2 cup plain, low-fat Greek yogurt
- 2 Tbsp. fresh lime juice
- 1 tsp. Dijon mustard
- Salt and freshly ground black pepper
- 8 (8-inch) whole-wheat tortillas

In mixing bowl, add all ingredients, and gently combine. Spoon 1/2 cup mixture onto bottom of each tortilla. Roll and serve.

Makes 8 servings.

Per serving (2 halves): 367 calories, 10 g total fat (1 g saturated fat), 58 g carbohydrate, 15 g protein, 10 g dietary fiber, 425 mg sodium.

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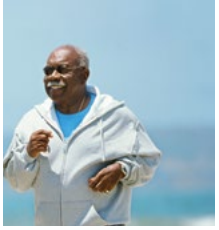
MAN 2 MAN

ASK THE DOCTOR

ELIAS OBEID, MD, MPH

I've heard about a test that might detect prostate cancer early called a PSA test. What is the PSA Test? Is this a test I should have?

Prostate Specific Antigen (PSA) is a protein produced by cells of the prostate gland. The test measures the level of PSA in a man's blood. The PSA blood level can be elevated in men with prostate cancer. There are other conditions that can cause a PSA level to rise, such as prostatitis (inflammation or infection of the prostate) or enlargement of the prostate called benign prostatic hyperplasia (BPH).

Recently, there has been discussion about the benefits of PSA screening in men without symptoms of prostate cancer. Some doctors think that this test can produce false-positive or false-negative results. A false-positive test result can occur when a PSA level is high but no cancer is found. This may cause anxiety and lead to other medical procedures such as biopsy. A false-negative test result can occur when a PSA level is low even though a man has prostate cancer.

Some organizations recommend that men who are at high risk of prostate cancer begin screening at age 40 or 45. High risk men include African American men and men whose first degree relative (father or brother) had prostate cancer. Other organizations still recommend yearly PSA testing and some do not. It is important to have a discussion with your doctor to determine if the PSA screening test is right for you.

Please email your "ask the doctor" questions to: rapinfo@fccc.edu. We cannot answer personal medical questions through this feature.

BREAST CANCER IN MEN

LISA BEALIN, BS

Although rare, it is important for men to remember that they can also develop breast cancer. Risk factors for male breast cancer can include obesity, a rare genetic condition called Klinefelter syndrome, and gynecomastia (excess breast tissue). A male with a BRCA mutation is also at a higher risk of developing breast cancer.

Symptoms of breast cancer in men are similar to those in women. Possible symptoms of male breast cancer include:

- **A lump or swelling which is usually (but not always) painless**
- **Skin dimpling or puckering**
- **Nipple retraction (turning inward)**
- **Redness or scaling of the nipple or breast skin**
- **Discharge from the nipple**

It is also important to know that not everyone with breast cancer will have symptoms. If you believe you are at risk to develop breast cancer, talk to your doctor who can do a clinical breast exam, to help with early detection. Your doctor may decide to order a mammogram, based on your risk factors or on clinical exam findings.

ELECTRONIC CIGARETTES:
PROCEED WITH CAUTION

MARGIE CLAPPER, PHD and ARLENE WARTENBERG, PHD

Tobacco contributes to 5 million deaths worldwide each year. For centuries, cigarettes have stayed the same: paper wrapped around tobacco. However, in 2004, Han Li, a Chinese pharmacist, helped develop the e-cigarette – and a $1.5 billion-a-year industry was born.

E-cigarettes (e-cigs) use a battery to heat liquid nicotine, making a vapor that can be inhaled. They can be purchased almost anywhere, including online and convenience stores. Many new "vaping" shops are opening to provide e-cig users a place to socialize. Manufacturers and advocates of e-cigs are actively promoting them through online social media and many unproven claims are in the news and advertisements. There are several hundred brands now on the market and their use, especially among youth, has increased greatly.

E-cigs are not controlled by the FDA (Food and Drug Administration), which raises concerns about the health risks and safety of this product. A December 2014 article in the New York Times discussed the lack of supervision in the production of e-cigs in China, the producer of 90% of the world's e-cigs. Experts report that "flawed or sloppy manufacturing" could account for dangerous heavy metals found in some e-cigs. Some Chinese factories have tried to monitor quality and safety, but others have not.

Nickel and chromium have been found at harmful levels (four times those of traditional cigarettes) in e-cigs as well as carcinogens and other dangerous compounds. Overheating of the lithium batteries or electronic chargers has led to the explosion of e-cigs, causing burns. Child poisoning cases from drinking liquid nicotine are also on the rise, including one death.

The large numbers of e-cigs on the market and the various chemicals in them make it very difficult to draw conclusions about their effect on health. Long exposure to the mist of propylene glycol, a major ingredient of e-cigs, should be avoided as it can affect the nervous system and cause irritation of the eyes and throat. E-cig use also narrows the peripheral airways, a concern for people with chronic lung disease. Similar to traditional cigarettes, nonsmokers are exposed to the vapors released from e-cigs as well as the nicotine that builds up on surfaces.

E-cigs have become very popular, especially among youth, because of their availability in hundreds of flavors. While the FDA says artificial flavors like strawberry, mint, and vanilla are safe in food, the health risks from breathing these flavors have not been tested. Even more worrisome is the attraction of the flavorful e-cigs to teenagers, 20% of whom have never smoked cigarettes before. Despite laws in 40 states that ban the sale of e-cigs to minors, use of e-cigs by youth (grades 6 -12) more than doubled between 2011 and 2012. Use of e-cigs among never-smoking young students increased 3 times between 2011 and 2013.

It is unclear if e-cigs can help smokers quit. A few studies suggest that e-cig use may help to stop or reduce smoking. However, the reported quit rates are much lower than those seen in studies looking at other commonly used smoking cessation therapies (such as Varenicline, a prescription medicine used to treat nicotine addiction). In addition, e-cig use could lead to increased nicotine use and dependence, and may serve as a gateway to traditional cigarettes. Also, a recent study shows that young e-cig users who have never smoked are more likely to try traditional cigarettes than youth who have never used e-cigs.

Cigarette packs come with a health hazards warning label–but this is currently not true for e-cigs. The FDA is considering regulations, but this will take time. In the meantime, Dr. Margie Clapper, co-leader of the Cancer Prevention and Control Program at Fox Chase Cancer Center, and her research team are working hard to study the long and short-term health effects of e-cigs.

MARK YOUR CALENDAR

FORCE (Facing Our Risk of Cancer Empowered) will hold their 9th annual "Joining FORCEs Against Hereditary Cancer" conference in Philadelphia on June 18-20, 2015. This is an international conference designed by and for people and families affected by hereditary cancer or a BRCA mutation.
Location: Philadelphia Downtown Marriott Hotel.
Information: www.facingourrisk.org

CANCERVENTION: a young adult with cancer conference: patient and caregiver program.
Saturday, April 18, 2015 from 9-4. The Hub Cira Center, Philadelphia. www.yacancerconnection.org/cancervention/

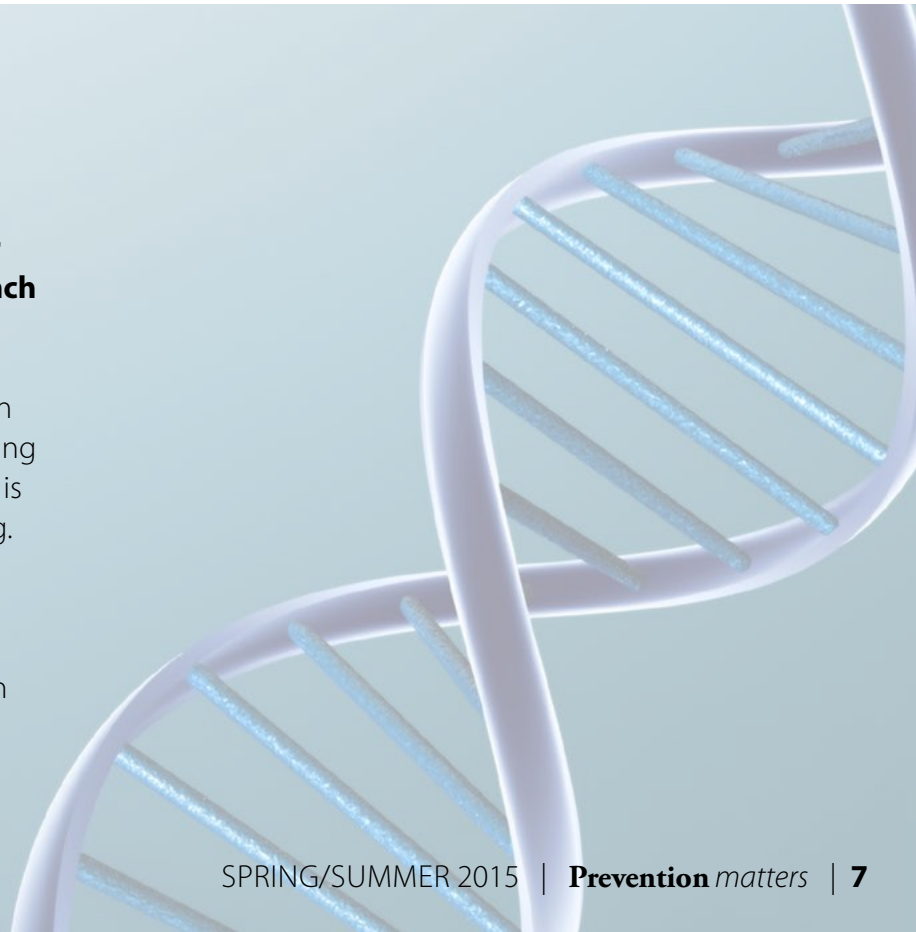
OPPORTUNITY TO ENROLL IN HEREDITARY CANCER RESEARCH

PROMPT (Prospective Registry of Multiplex Testing) is an online research registry for patients and families who have had multigene panel testing, and who have been found to have a genetic mutation or variant of uncertain significance. It provides patients, doctors, and researchers an opportunity to share information about panel testing. Patients can enroll in PROMPT and provide data by completing questionnaires about their personal and family health histories.

PROMPT is a joint effort with several academic medical centers and commercial labs, working together to learn more about the genes that are studied on multigene panels. If you register, you will be asked to complete some online surveys. Fox Chase Cancer Center is not directly involved with this study. To learn more, please visit the study website at www.promptstudy.org.

OPPORTUNITY TO ENROLL IN A PREVENTION STUDY FOR LYNCH SYNDROME PATIENTS

"Naproxen in Preventing DNA Mismatch Repair Deficient Colorectal Cancer in Patients With Lynch Syndrome" is a National Cancer Institute sponsored clinical trial for patients who have Lynch syndrome. This clinical trial will study whether chemoprevention with the drug naproxen may keep cancer from forming in patients with Lynch syndrome. Chemoprevention is the use of certain drugs to keep cancer from forming. 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. Fox Chase Cancer Center is not directly involved with this study. To learn more about this study, visit the National Cancer Institute Clinical Trials website at https://clinicaltrials.gov/ct2/show/NCT02052908



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The Department of Clinical Genetics offers the most comprehensive risk assessment program in the Philadelphia region. It encompasses all of Fox Chase Cancer Center's clinical services for people at risk for cancer, as well as innovative research in the areas of cancer prevention and genetics.

CONTACT THE RISK ASSESSMENT PROGRAM:
1-877-627-9684 | rap-info.fccc.edu

Mary Daly, MD, PhD
Chair
Department of Clinical Genetics

Michael Hall, MD, MS
Director
Gastrointestinal Cancer Risk

Hossein Borghaei, DO
Director
Lung Cancer Risk

Elias Obeid, MD, MPH
Assistant Professor
Breast, Ovary, and
Prostate Cancer Risk

SURVEY SAYS

From August to November 2014 we asked our readers to complete a brief survey about our newsletter, Prevention Matters. Thank you to everyone who responded (the survey is now closed). 842 people completed the survey.

THE SURVEY RESULTS:

- Our readers want the newsletter published twice a year. Many readers prefer receiving it by email.
- Readers like the content, reading level and length of newsletter.
- The sections read most include research study updates, cover feature stories, and cancer patient stories. Also popular are the genetic counselor columns and recipes.
- Readers want to read more about healthy living (nutrition, exercise); new research studies to join; updates on cancer treatment and survival; genetics and family history, and information resources such as events and links to journal articles.

Please continue to send your comments or questions to rapinfo@fccc.edu . We welcome your suggestions.

Editors: Lisa Bealin, Yana Chertock, Nina Galpern and Joann Sicilia **Designed by 4545 Creative Services**

Risk Assessment Program
333 Cottman Avenue
Philadelphia, PA 19111-2497

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