The Johns Hopkins Guide for
Patients and Families:

Peutz-Jeghers Syndrome

JOHNS HOPKINS
MEDICINE

The Johns Hopkins Guide for
Patients and Families

Peutz-Jeghers Syndrome

THE JOHNS HOPKINS GUIDE FOR PATIENTS AND FAMILIES:
PEUTZ-JEGHERS SYNDROME

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INTRODUCTION

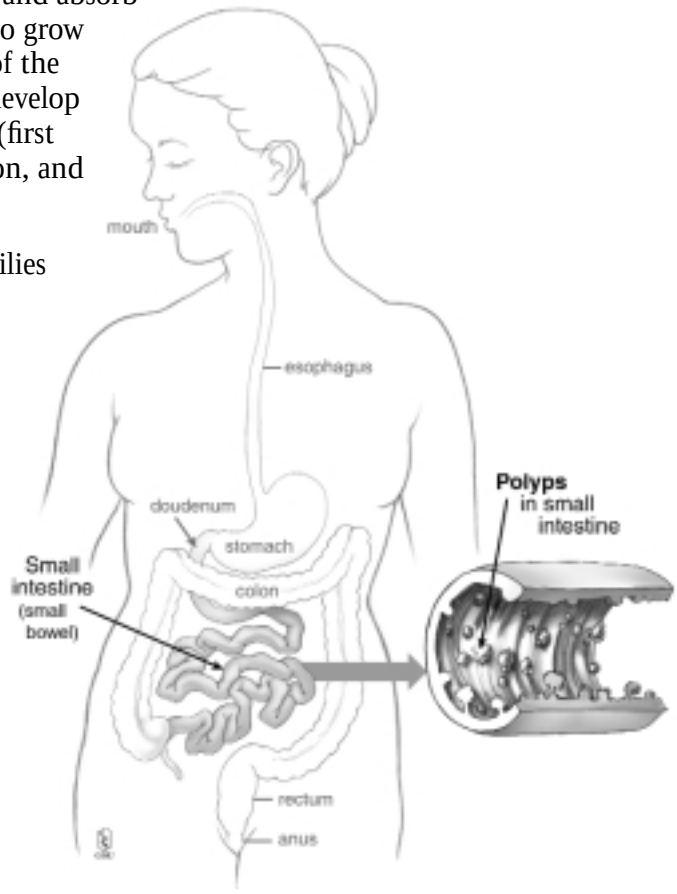
The purpose of this booklet is to help you understand Peutz-Jeghers Syndrome (PJS). PJS is a condition that tends to run in families and increases the risk for developing cancer, especially colon cancer. The information in this booklet is intended to add to, and not replace, discussions with doctors, genetic counselors, nurses, and other members of your health care team. We suggest that you read the booklet all the way through in the order it was written, as each new section builds upon information in previous sections. Words in **bold print** are defined in the glossary at the end of this booklet on page 15.

Remember, anyone who already has cancer needs to continue receiving regular and thorough medical check-ups. However, it is just as important for some family members who may be at risk for cancer to have regular check-ups. For more information, please contact your health care provider or the Hereditary Colorectal Cancer Registry at The Johns Hopkins Hospital at (410) 955-3875 or toll free 1-888-77-COLON (1-888-772-6566).

WHAT IS PEUTZ-JEGHERS SYNDROME?

Peutz-Jeghers Syndrome (PJS) is a rare condition that tends to run in families. About 1 in 160,000 to 1 in 280,000 persons will develop PJS. It is caused by a change (**mutation**) in a **gene** that increases the risk for developing **colon** and other cancers. People with PJS develop **polyps** in the digestive tract (**gastrointestinal (GI) tract**). A polyp is a growth that sometimes takes the shape of a mushroom and varies in size from less than 1/10 of an inch to 1 to 2 inches. The GI tract (as seen below) is a hollow tube that begins at the mouth and ends at the anus. It has several parts including the esophagus, stomach, small intestine (small bowel) and large intestine (colon). The function of the GI tract is to digest food and absorb the nutrition your body needs to grow and remain healthy. The areas of the GI tract that most commonly develop polyps include the **duodenum** (first part of the small intestine), colon, and stomach.

Other cancers can occur in families with PJS, including esophagus, stomach, small intestine, colon, pancreas, lung, and kidney. Women have a higher risk for cancers of the breast, ovaries, uterus (**endometrium**), and rarely, cancers of the cervix. Men, especially young boys, have a higher risk of getting tumors of the testicles.



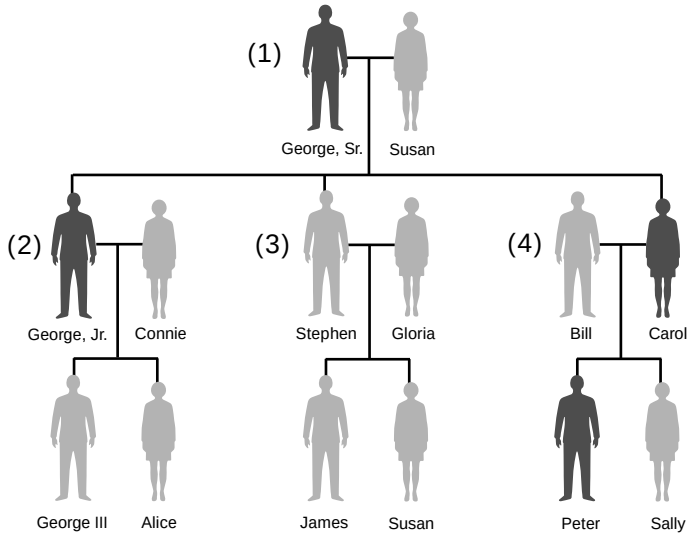
HOW IS PJS INHERITED?

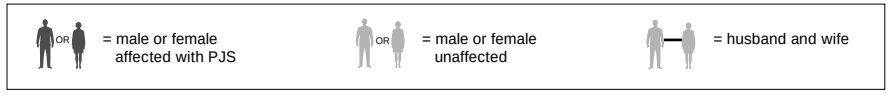
Scientists are studying which gene mutations may cause PJS. Some people with PJS carry a mutation in a gene called **STK 11**, also known as LKB1, which is located on **chromosome** 19. Usually, they get this mutated gene from one of their parents. People with PJS have a 50% chance of passing on the mutation to each of their children. The family tree shown on the next page is an example of how PJS can be passed on in a family.

Not all people with PJS have a family history of PJS. Some may be the first in their family to get PJS, and are believed to have a new mutation. They, too, can pass this new PJS gene mutation on to their children.

Genetic counseling is available, and recommended, for people with PJS and their family members. Genetic counselors explain how PJS is inherited and discuss which family members are **at-risk** for developing the condition. Genetic counseling services are becoming increasingly available through genetics and oncology (cancer) departments in many hospitals. It is recommended that families with PJS contact local registries (see resources on pages 11 through 13) for educational and support resources to help them identify at-risk family members and keep up-to-date on the latest knowledge about this disease.

EXAMPLE OF A FAMILY WITH PJS





(1) George, Sr. has PJS. His wife, Susan, is unaffected (this means she does not carry the PJS gene). They have three children, George, Jr., Stephen and Carol. Because George, Sr. has PJS, each of his children had a 50% chance of getting PJS. In this case, George, Jr. and Carol inherited PJS (shown by the darkened figures).

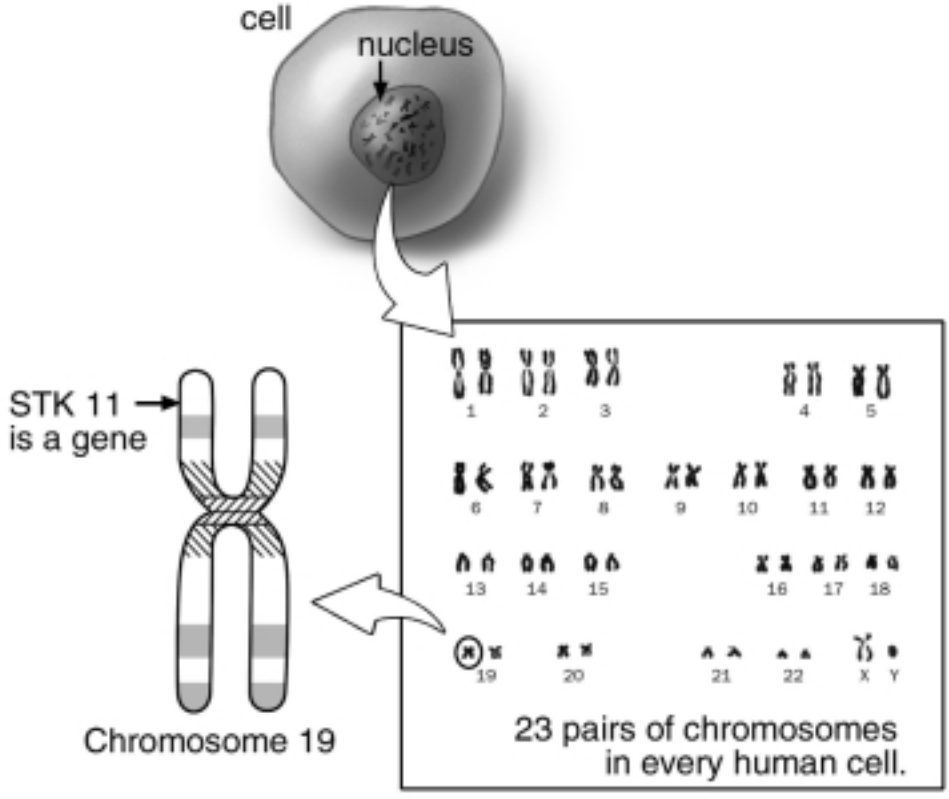
(2) George, Jr., and his wife, Connie, have two children. Although both children had a 50% chance of getting PJS from their dad, neither did.

(3) Stephen and his wife, Gloria, have two children. Since Stephen and Gloria are both unaffected, there was no risk of passing PJS on to their children.

(4) Carol and her husband, Bill, have two children. Both children had a 50% chance of getting PJS from their mother, but only Peter got PJS.

IS THERE A BLOOD TEST TO IDENTIFY PEOPLE WITH PJS?

There is a blood test available commercially that can find the mutated STK 11 gene causing PJS in most patients. A genetic counselor can help to determine if gene testing is right for you and your family members. See “Additional Resources” on pages 11 through 13 to find a genetic counselor near you.



WHAT ARE THE SYMPTOMS OF PJS?

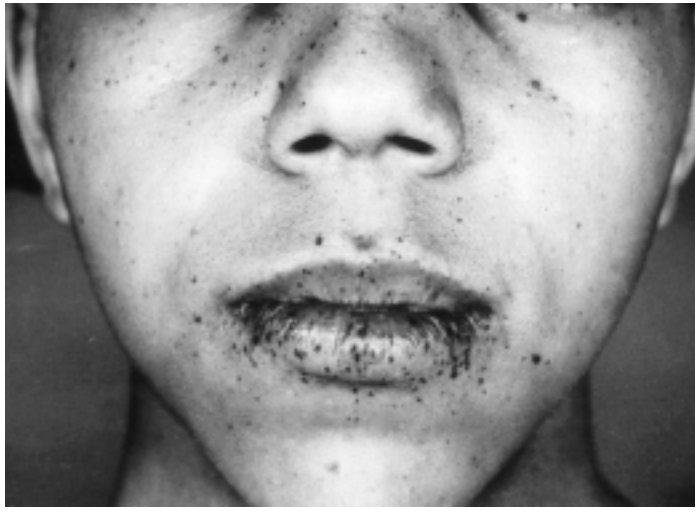
Each person with PJS may have different combinations of the symptoms discussed here.

Polyps: People with PJS often develop a unique type of polyps called **hamartomas**. These occur mostly in the small intestine but can also be found in the stomach and/or colon. They usually begin forming during childhood, but symptoms often do not develop for years. A major concern in PJS is that a hamartoma can cause bleeding that may or may not be seen in the stool. Rarely, hamartomas can develop into cancer.

Freckling: Most people with PJS have a distinct freckling pattern (called **melanin spots**) on certain parts of their body (see picture below). They usually form in early childhood, but may develop later. They can fade during the late teenage years. They are completely harmless. Melanin spots look like small, flat, dark brown or black freckles on the lips and inner cheeks. Sometimes they show up on the eyelids, hands, and feet.

GI Problems: Diarrhea (not as a result of diet or flu), constipation, crampy pain and/or bloating in the belly, weight loss, lack of energy, anemia (low red blood cell count), nausea, and precocious (early) puberty can occur with PJS. Occasionally, men develop excessive breast tissue. Sometimes, children and adults may develop sudden, severe abdominal pain caused by polyps blocking the intestine called **intussusception**. This is an emergency that requires prompt medical attention.

Example of Melanin Spots in PJS



There is no safety in waiting for symptoms to develop. It is vital that parents and guardians make every effort to have at-risk children start screening during the teenage years even if they do not have symptoms.

WHY IS EARLY DIAGNOSIS IMPORTANT?

Early diagnosis of PJS is important for the prevention and/or early detection of cancer. Not everyone who has PJS may get cancer, but PJS increases cancer risk. A blood test for the PJS gene can tell a person if he or she has received the gene mutation found in the family, or it can confirm a suspected diagnosis of PJS. It is helpful to know which relatives have inherited PJS because they have a higher risk of getting cancer at a younger age compared with the general population. When cancer is caught early, the chance of curing it is much better. Therefore, finding signs or symptoms of PJS early can greatly reduce the risk for cancer. The following screening tests are recommended on a regular basis for individuals at risk for PJS. These tests were selected for the Johns Hopkins Hospital based on experience and scientific literature, but may vary between institutions.

Age (year)	Procedure
From birth to age 24	Children and adults should have a history and physical exam every year with routine blood tests. The doctor will look for melanin spots, precocious puberty, and testicular tumors.
By age 12	Parents should discuss genetic testing (STK 11 gene) for their child if possible. Otherwise, as below.
At ages 12, 18, and 24	Children and adults should have an upper endoscopy[1], colonoscopy[2], and small bowel series[3] at each of these ages.

1. **Upper endoscopy** looks at the upper part of the GI tract for polyps and cancer using a thin, flexible, lighted tube that is inserted through the mouth. A small sample of tissue can be removed (**biopsy**) and viewed under a microscope for abnormalities. A local **anesthetic** is sprayed in the throat before the test to minimize discomfort. A light anesthesia will make you sleepy during the test. It is important not to eat or drink after midnight before this test unless your doctor tells you otherwise.
2. **Colonoscopy** looks for polyps and signs of cancer along the entire colon using a thin, flexible, lighted tube that is passed through the rectum. A light anesthesia will make you sleepy during the test. You will be provided with instructions to prepare for this test. Please follow them carefully to ensure good test results.
3. A **small bowel series** takes several X-rays of your small intestine to look for the cause of intestinal blockages such as tumors, strictures (narrowing of any tube in the body), inflammation (tissue that is swollen, red, and/or irritated), and diseases of the small intestine. After you drink a **barium** milkshake through a straw, a special X-ray called a fluoroscope takes a series of pictures as the barium flows through the small intestine. This test usually takes between 1 to 2 hours depending on how fast the barium travels. An upper GI series, which takes X-ray pictures of the lower esophagus, stomach, and duodenum, is often performed at the same time.

HOW IS THE DIAGNOSIS FOR PJS MADE?

The diagnosis of PJS is made if a person has polyps in the GI tract and at least two of the following: polyps in the small bowel, melanin spots, and/or a family history of PJS.

If all exams are normal through the age of 24, an at-risk person probably does not have PJS. If the at-risk person undergoes genetic testing and is negative for the PJS gene mutation found in an affected family member, then the individual can follow screening recommendations for the general population (see the *appendix* on pages 17 and 18). If polyps are detected, the individual should begin the surveillance recommendations on page 9.

WHAT KIND OF FOLLOW-UP CARE IS NEEDED FOR PJS?

If polyps are found during an upper endoscopy or a colonoscopy, they should be removed. Alternatively, the doctor may recommend a **laparotomy**, a surgical procedure in which an incision is made in the belly or pelvic region and polyps are removed.

If *cancer* is found during a test, the patient and doctor will discuss the best course of action, which may include more extensive surgery and, perhaps, additional treatment such as chemotherapy.

Finding and removing polyps may result in the prevention of cancer in the colon or small bowel, and in a longer life. However, people with PJS are still at higher risk for cancers of the stomach, pancreas, lungs, kidneys, breast, ovaries, uterus (endometrium), cervix, and testicles. There are ways to screen and monitor people with PJS to help detect these cancers at early stages. On the next page are recommendations for regular follow-up of individuals with PJS based on what is presently known by experts in cancer and genetics. Again, these procedures were selected for the Johns Hopkins Hospital based on experience and scientific literature, but may vary between institutions. Persons with PJS should contact their physicians regularly for any new information. In addition, counseling may be helpful to deal with the stress of frequent screenings and procedures.

Surveillance Recommendations for People with PJS

Age (years)	Procedure for Males	Procedure for Females
From 12 on	History and physical exam (precocious puberty and testicular tumors) every year with routine blood tests	History and physical exam (precocious puberty) every year with routine blood tests
	Upper endoscopy, colonoscopy and small bowel series every 2 years	Upper endoscopy, colonoscopy and small bowel series every 2 years
From 18 on	*Continue above*	*Continue above, plus:* Gynecological exam, transvaginal ultrasound[1], and CA 125[2] every year
From 25 on	*Continue above, plus:* **CT scan** every 1 to 2 years OR Endoscopic ultrasound[3] and CA 19-9[4] every 1 to 2 years	*Continue above, plus:* CT scan every 1 to 2 years OR Endoscopic ultrasound[3] and CA 19-9[4] every 1 to 2 years. Breast exam and **mammogram** every year

1. **Transvaginal Ultrasound** is a technique to find ovarian cancer and other masses by bouncing sound waves off the ovaries to create pictures called sonograms.
2. **CA 125** is a **tumor marker** for ovarian cancer that is measured in a sample of blood drawn from the arm. It is, however, not always present in women with ovarian cancer, and it is sometimes present in women who do not have ovarian cancer. For this reason, a gynecological exam, transvaginal ultrasound, CT scan, and mammogram may also be recommended.
3. **Endoscopic Ultrasound** is a picture-taking technique used to examine the pancreas since people with PJS have a higher risk of developing pancreatic cancer. It is performed by inserting a flexible tube through the mouth and into the stomach. The endoscope has an ultrasound wand which allows detailed pictures of the pancreas to be taken. A local anesthetic is sprayed in the throat before the test to minimize discomfort. A light anesthesia will make you sleepy during the test. It is important not to eat or drink after midnight before this test unless your doctor tells you otherwise.
4. **CA 19-9** is a tumor marker for pancreatic cancer that is measured in a sample of blood drawn from the arm. Tumor markers are biochemical substances in the blood that, if found to be at higher than normal levels, may suggest the presence of cancer.

SUMMARY

Peutz-Jeghers Syndrome is a rare condition that tends to run in families. It is caused by a gene mutation. Symptoms can include polyps in the GI tract and melanin spots on the lips, eyelids, hands, and feet. PJS increases the risk for developing cancer, most commonly of the breast and colon. Starting regular screening for PJS and cancer at an early age can ensure early diagnosis of disease. Currently, a gene test for PJS is available. Ask your health care provider for the latest information on PJS to take full advantage of any new screening or testing methods.

ADDITIONAL RESOURCES

1. THE JOHNS HOPKINS COLON CANCER RISK ASSESSMENT SERVICE

Johns Hopkins Outpatient Center, 8th floor
Johns Hopkins Hospital
601 North Caroline Street
Baltimore, Maryland 21287
Phone (410) 614-LIFE (410-614-5433)

Offered through the Johns Hopkins Oncology Center, this service provides the opportunity to meet with medical experts to review family medical history, risk assessment, genetic counseling and testing, and recommendations for screening, prevention, and detection. Families with PJS will receive specific recommendations through a consultation with this service.

2. POLYPOSIS AND HEREDITARY COLON CANCER REGISTRIES

Registries located throughout the United States, Canada and in other countries study families with PJS. Registries can help to identify at-risk relatives, and may be able to provide educational materials and physician referrals. For more information, contact:

Coordinator, Hereditary Colorectal Cancer Registry
The Johns Hopkins Hospital
550 North Broadway, Suite 108
Baltimore, Maryland 21205-2011
Phone (410) 955-3875
Toll free 1-888-77-COLON (1-888-772-6566)
Fax (410) 614-9544
E-mail hccregistry@jhmi.edu
http://www.hopkins-coloncancer.org

3. THE MID-ATLANTIC CANCER GENETICS NETWORK (MACGN)

MACGN is a regional center of the national Cancer Genetics Network and a resource for patient education materials on a variety of hereditary and familial cancers. If you are concerned about your risk for Peutz-Jeghers Syndrome or cancer because of a family history, please call MACGN for more information or a referral at toll free 1-877-880-6188 or visit the Web site at http://www.MACGN.org.

4. CANCER INFORMATION SERVICE (CIS)

Toll free 1-800-4-CANCER (1-800-422-6237)

The Cancer Information Service (CIS) is a national toll free telephone inquiry system that provides information about cancer and cancer-related resources to the general public, patients and their families, and health professionals. Most CIS offices are associated with Comprehensive Cancer Centers or community hospitals. CIS counselors provide support, understanding, and rapid access to the latest information, as well as referrals to local services and resources. They do not diagnose or give treatment recommendations. All calls are confidential and callers need not give their names.

5. AMERICAN CANCER SOCIETY (ACS)

National Headquarters
1599 Clifton Road, N.E.
Atlanta, Georgia 30329
Phone (404) 320-3333
Toll free 1-800-ACS-2345
http://www.cancer.org

The ACS can offer assistance and cancer resources. Check the telephone directory for your local chapter.

6. GENERATION TO GENERATION

A newsletter for people concerned about hereditary colon cancer and polyposis, this publication is free to all individuals and their families with these conditions. The editors welcome suggestions, questions, and personal stories. Articles concerning new research, cancer genetics and treatments are highlighted. To subscribe, contact:

Editor, Generation To Generation
Department of Behavioral Science
Box 243
M. D. Anderson Cancer Center
1515 Holcombe Boulevard
Houston, Texas 77030

7. STATE VOCATIONAL REHABILITATION SERVICE

This service offers training for another vocation if one should be physically unable to return to the same kind of work performed prior to surgery. Check your telephone directory for your state's services.

8. NATIONAL ORGANIZATION FOR RARE DISORDERS (NORD)

PO Box 8923
Fairfield, CT 06812
Toll free (800) 999-6673
General (203) 746-6518

A national helpline for rare disorders. The automated message system will link people with the appropriate help for their disorder. To speak to a live person, call the general number listed.

9. GENETIC ALLIANCE

4301 Connecticut Avenue, NW, #404 Washington, DC 20008-2304
phone (202) 966-5557
Helpline only (800) 336-GENE
http://www.geneticalliance.org
Genetic Alliance is an international coalition of individuals, professionals and genetic support organizations working together to enhance the lives of everyone impacted by genetic conditions.

10. THE PEUTZ-JEGHERS SYNDROME ON-LINE SUPPORT GROUP

http://listserv.acor.org/SCRIPTS/WA-ACOR.EXE?SUBED1=pjs&A=1

This patient-run on-line support group is part of the Association for Cancer Online Resources (ACOR). ACOR is a large collection of cancer-related Internet resources. In addition to supporting the mailing lists, ACOR develops state-of-the-art Internet-based knowledge systems that allow the public to find and use credible information relevant to their illness. Also links to inherited cancer registries.

11. THE NATIONAL CANCER INSTITUTE'S CANCER GENETICS SERVICES DIRECTORY

http://cancernet.nci.nih.gov/genesrch.shtml

The Cancer Genetics Services Directory is part of the National Cancer Institute's CancerNet Web site. The directory allows you to search for providers by type of cancer or cancer syndrome, location, and name.

12. THE NATIONAL SOCIETY OF GENETIC COUNSELORS (NSGC)

The NSGC can help you find a genetic counselor near you.
http://www.nsgc.org.

SUGGESTED READING FOR ADDITIONAL INFORMATION:

Taking Time: Support for People with Cancer and the People Who Care About Them, National Cancer Institute. Call CIS at 1-800-4-CANCER.

What You Need to Know about Cancer of the Colon and Rectum, National Cancer Institute. Call CIS at 1-800-4-CANCER.

Understanding DNA Testing: A Basic Guide for Families, National Center for Education in Maternal and Child Health, 38th and R Street NW, Washington, DC 20057.

GLOSSARY

Anesthetic - An oral or injected medicine used to reduce discomfort during medical procedures by producing local or general insensibility to pain through numbness or sleep.

At-risk - A person "at-risk" has the possibility of developing the condition that is present in his/her family.

Barium - A chalky liquid that is visible on X-ray film and permits the doctor to see any defects, obstructions, or masses.

Biopsy - The removal of a sample of tissue that is then viewed under the microscope.

CA 19-9 - A tumor marker for pancreatic cancer found in the blood that, if elevated, suggests the presence of cancer.

CA 125 - A tumor marker for ovarian cancer found in the blood that, if elevated, suggests the presence of cancer.

CAT or CT Scan - (Computerized Axial Tomography) A form of X-ray that shows the size and shape of body organs layer by layer.

Cecum - The first part of the large intestine.

Chromosome - The carrier of the genetic material (genes) of a cell. The normal number of chromosomes in a human cell is 46 (23 pairs).

Colon - The last portion of the intestines. It comprises the cecum, ascending colon, transverse colon, descending colon, and sigmoid colon. It is 5-6 feet in length. Also known as the large intestine or large bowel.

Colonoscopy - A test in which a flexible tube, about 5 feet in length, is used to examine the inside of the entire colon. The tube has a light source, a magnifying eyepiece, and an open channel through which air can be passed and a biopsy taken.

DNA - (Deoxyribonucleic Acid) The molecule that contains the code for the genetic blueprint. It is found in the nucleus of cells.

Double Contrast Barium Enema - A test in which a chalky substance called barium sulfate and air are used to expand the colon so that X-rays of the colon can be taken.

Duodenum - The first part of the small intestine. It is 12-15 inches in length.

Endometrium - The mucous membrane comprising the inner layer of the womb (uterus).

Fecal Occult Blood Test - A test using specially treated cardboard slides to check for hidden blood in the stool.

Flexible sigmoidoscopy - A test in which a flexible tube about 2 feet in length is used to examine the rectum and lower part of the large bowel. The tube has a light source, a magnifying eyepiece, and an open channel through which air can be passed and a biopsy taken.

GI tract - (Gastrointestinal tract) The digestive system, consisting of the esophagus, stomach, small intestine (22-25 feet in length), and large intestine (5-6 feet in length).

Gene - The basic unit of heredity, each one occupying a certain place on a chromosome.

Hamartoma - A type of polyp that is made of an abnormal mixture of tissue types but is not cancerous.

Hereditary - Genetically passed down from parent to child.

Ileum - The third and longest portion of the small intestine about 12 feet long.

Intussusception - A blockage of the intestines.

Jejunum - The middle portion of the small intestine in between the duodenum and ileum, about 8 feet long.

Laparotomy - A surgical procedure in which an incision is made in the abdomen and polyps are removed; requires full sleep anesthesia.

Mammogram - An X-ray of the breast.

Melanin - Skin pigment.

Melanin spots - Dark, pigmented spots that can occur on the lips, mouth, fingers, toes, eyelids, and genitals of individuals with PJS.

Mutation - A change in a gene that may result in a specific disorder.

Oncology - The study of tumors. The name of the hospital department where a cancer patient frequently goes for treatment.

Pap Test - A test which involves taking some cells from a woman's cervix and looking at them under a microscope to see if abnormal or cancer cells are present. Also called a Pap smear.

Polyp - An abnormal, flat, raised or mushroom-shaped growth.

Peutz-Jeghers Syndrome - An inherited disorder of the gastrointestinal tract in which there are polyps that can become cancerous. The polyps occur mostly in the small intestine (small bowel) but can also develop in the stomach and colon. In PJS there are also skin pigment changes.

PSA - (Prostate Specific Antigen) A tumor marker for prostate cancer found in the blood that, if elevated, suggests the presence of cancer.

STK 11 - Mutation of this gene is found in patients affected with PJS.

Syndrome - A collection of abnormal physical characteristics occurring in an individual (Example: Peutz-Jeghers Syndrome has polyps in the GI tract and occasional freckling of the lips or mouth).

Transvaginal ultrasound - (TVS) A test for ovarian tumors or cancer that bounces sound waves off the ovaries to create pictures called sonograms.

Tumor marker - A substance sometimes found in an increased amount in the blood, other body fluids, or tissues that may suggest the presence of some types of cancer.

Ultrasound - A test that bounces sound waves off tissues and internal organs and changes the echoes into pictures called sonograms. Tissues of different densities reflect sound waves differently.

Upper Endoscopy -
(Esophagogastroduodenoscopy, EGD) A test in which a flexible tube is used to examine the interior of the upper GI tract (esophagus, stomach, and duodenum). The tube has a light source, a magnifying eyepiece, and an open channel through which a biopsy can be taken.

APPENDIX:
CANCER SCREENING GUIDELINES FOR THE GENERAL POPULATION

The American Cancer Society (ACS) recommends the following guidelines for early detection of cancer in people without symptoms or a family history of cancers:

Cancer-related checkup every 3 years for people age 20-40 and every year if over 40. This exam should include the procedures listed below and health counseling (such as tips on quitting smoking) and, depending on the person's age, might include tests for cancers of the thyroid, oral cavity, testicles, prostate, ovaries, skin, and lymph nodes.

Breast

- Self-exam every month for women age 20 or over
- Clinical breast exams by doctor every three years for women age 20-39, every year if over 40 (This exam should be done close to the time of the scheduled mammogram.)
- Mammogram every year for women age 40 or over

Female Reproductive System

Pelvic exam and **PAP test** every year for women who are, or who have been, sexually active or have reached age 18; after 3 or more consecutive satisfactory normal exams the PAP test may be performed less frequently at the doctor's discretion.

Colon/Rectum

People age 50 or over without any risk factors for colorectal cancer

- **Flexible Sigmoidoscopy** every 5 years
  OR
- **Fecal Occult Blood Test** (FOBT) every year
  OR
- **Flexible Sigmoidoscopy** every 5 years AND **FOBT** every year
  OR
- **Colonoscopy** every 10 years
  OR
- **Double Contrast Barium Enema** every 5 years

The ACS prefers flexible sigmoidoscopy every 5 years together with annual FOBT compared with flexible sigmoidoscopy or FOBT alone since evidence shows that the two methods together are more effective than either one alone.

CANCER'S SEVEN WARNING SIGNALS

If you have any of the *Seven Warning Signals* listed below, see your doctor or go to your clinic without delay.

1. Change in bowel or bladder habits

2. A sore that does not heal

3. Unusual bleeding or discharge

4. Thickening or lump in breast or elsewhere

5. Indigestion or difficulty in swallowing

6. Obvious change in wart or mole

7. Nagging cough or hoarseness

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To order copies of this booklet, please contact the Coordinator, Hereditary Colorectal Cancer Registry, The Johns Hopkins Hospital, 550 North Broadway, Suite 108, Baltimore, MD 21205, 1-888-77COLON (772-6566), e-mail: hccregistry@jhmi.edu.