Familial Ovarian Cancer Registry

at Roswell Park

ROSWELL PARK
COMPREHENSIVE CANCER CENTER

PATIENT EDUCATION

The Familial Ovarian Cancer Registry is a collection of DNA samples, family history profiles and lifestyle information from patients and families with a history of ovarian cancer. Researchers use this collection to monitor patterns and trends in the disease; identify new genes associated with ovarian cancer; and develop new ways to detect, treat and prevent ovarian cancer.

Our registry, with information from nearly 3,000 families, is the largest such database in the world. In addition to our own research teams here at Roswell Park, we provide access to the Registry to top scientists from around the globe to support and expand critically needed study of ovarian cancer.

Familial Ovarian CANCER REGISTRY

Q&A on Ovarian Cancer

These are the most frequent questions asked about ovarian cancer at the Familial Ovarian Cancer Registry.

What is ovarian cancer?

There are almost 40 different types of ovarian cancers. However, nine out of 10 ovarian cancer patients have **epithelial** tumors, which begin in the tissue on the surface of the ovary (epithelium). These are called **adenocarcinomas** – a malignant (cancerous) tumor of epithelial origin which begins in glandular tissue.

Serous adenocarcinoma is seen most often, followed by **endometrioid**, **mucinous** and **clear cell** adenocarcinomas.

Carcinomas of **borderline malignancy** are a subgroup of serous and mucinous adenocarcinomas, which are usually less aggressive and have a significantly higher cure rate than serous and mucinous adenocarcinomas.

What causes ovarian cancer?

The specific cause(s) of ovarian cancer is unknown. What is known, however, is that affluent women in the world's most affluent countries have the highest rates – implying that something in these mostly industrialized countries may be a cause. What behaviors are more common among these women than women in poorer countries? There are several hypotheses:

- eating a high-fat diet
- using talcum powder on sanitary napkins or on the vaginal area
- infertility
- fertility drugs (**Clomid** and **Perganol**)

However, none of these has been confirmed.

Probably the only fact known to be associated with a significant increase in ovarian cancer is when women have two or more first- or first- and second-degree relatives with ovarian cancer or early-onset breast cancer.

A woman's lifetime chance of developing breast and/or ovarian cancer is greatly increased if she inherits an altered BRCA1 or BRCA2 gene. Women with an inherited alteration in one of these genes have an increased risk of developing these cancers at a young age (before menopause), and often have multiple close family members with the disease. Another condition associated with increased risk of ovarian cancer is hereditary nonpolyposis colorectal cancer (HNPCC), also known as Lynch syndrome.

What are the symptoms?

Ovarian cancer was believed to be a silent killer because people thought that by the time a woman had symptoms, the disease had already spread throughout her abdomen and beyond. It is now known that the early stages (I and II) are not universally silent and have the same symptoms as advanced stages (III and IV). Symptoms most often associated with ovarian cancer include: a feeling of being bloated; clothes that don't fit quite as easily as they once did; vague abdominal and pelvic discomfort; and gastrointestinal symptoms such as gas, back pain, and fatigue.

Although many women have these symptoms, if they persist for several weeks, they **could** be an early warning of ovarian cancer.

How is it diagnosed?

The only definitive way to diagnose ovarian cancer is surgery to remove the tumor for laboratory evaluation. Fortunately, there are tests to help determine if surgery is needed. In addition to a **pelvic exam, pelvic and vaginal ultrasound** of the ovaries can often (but not always) help distinguish between **malignant** (cancerous) and **benign** (noncancerous) tumors. Cystic tumors (i.e., no solid areas suggesting cancer) are usually benign. When solid areas or septation are seen on ultrasound, the chance of cancer increases.

CA125 levels (a tumor marker in the blood), which are elevated in eight out of 10 women with advanced (stage III and IV) disease and in one out of two women with cancer localized in the ovary (stage I), can be determined by a simple blood test. However, **CA125** levels can also be elevated in benign conditions such as endometriosis, pelvic inflammatory disease of the tubes and ovaries, uterine fibroids, pregnancy, and sometimes in cancer of the pancreas and of the gastrointestinal tract.

What does staging of ovarian cancer mean?

Stage refers to how far the disease has advanced. Accurate staging is important in treatment planning because the **prognosis** (outcome) worsens as the stage increases. Generally, there are four stages of ovarian cancer:

Stage I: cancer is limited to the ovary

Stage II: cancer has spread beyond the ovary but is still limited to the pelvis (below the navel)

Stage III: cancer has spread beyond the ovary but is still limited to the pelvis and abdominal cavity (excluding the liver)

Stage IV: cancer has spread to the liver or outside the abdomen, often involving the space surrounding the lung

What is the role of surgery?

Surgery is needed for **all stages** of ovarian cancer. If the cancer is **limited to the ovary,** surgery may be the only treatment needed. To document this, four areas within the abdominal cavity are evaluated:

- the undersurface of the diaphragm
- the omentum (a fatty apron that hangs down from the colon)
- lymph nodes along the abdominal aorta
- pelvic lymph nodes.

The abdominal cavity is also **washed** with a saline solution and the cells are stained to identify floating cancer cells not visible to the naked eye. For stages II, III, and IV, surgery is often performed before or in between chemotherapy. Surgery performed by a gynecologic oncologist, and removal of most of the tumor, results in the best survival.

What is the role of chemotherapy?

The most important **chemotherapy** (drug treatment) agents for ovarian cancer are Platinum compounds and Taxanes. These medications are usually given **intravenously** (through a vein) every three to four weeks, for six treatments. Patients are evaluated at each treatment and have a **CA125** test and blood work. If the **CA125** level was elevated before and is falling during chemotherapy, the treatment is almost certainly effective. If the **CA125** level rises significantly during chemotherapy, it

usually means that the treatment is not effective. Some women receive chemotherapy **intraperitoneally** (through a small catheter inserted into their abdominal [peritoneal] cavity). Intraperitoneal chemotherapy is often used when only very small deposits of cancer remain within the abdominal cavity after primary surgery.

If initial chemotherapy fails, what effective treatments are available?

There are other promising chemotherapy drugs available. **Topotecan**, **Gemcitabine**, **Tamoxifen**, **Doxil** or **oral etoposide** are effective in some women.

In some patients, newer agents such as **Avastin** (an anti-angiogenic agent) are used to cut off blood supply to the tumor.

Additionally, personalized medicine (testing and profiling the tumor for molecular and genomic changes) allows us to better develop targeted therapies that are effective for each patient's specific cancer.

What is the prognosis and outlook?

For those women diagnosed with ovarian cancer limited to the ovary (stage I), over 90 percent will be alive at five years. This contrasts dramatically with approximately 25 percent for those women diagnosed with stage III or IV ovarian cancer. Clearly, early detection and prompt diagnosis and staging with improved tests is a hope for the future.

What about early detection?

At the present time, there are no reliable early detection tests for ovarian cancer. Although CA125 could be elevated in ovarian cancer patients, in a significant number of patients the test may also be falsely positive or falsely negative. For women with a family history of ovarian cancer, the Registry does recommend cancer screening and surveillance that should include pelvic and abdominal examination,

CA125 blood levels and transvaginal ultrasound every six months, beginning between 25 and 35 years of age. Genetic consultation is recommended for any individual concerned about risk for ovarian cancer due to family history.

Are there any new strategies for early detection?

As stated before, at the present time, there are no reliable early detection tests for ovarian cancer. Therefore, Roswell Park Cancer Institute is developing new strategies for early detection.

What novel treatment strategies are available?

Advances in personalized medicine have led to the identification of several new targets that could be used for treatment of ovarian cancer. The strategies that are evolving include **anti-antiogenesis**, the use of targeted antibodies and other immunotherapies.

At Roswell Park, we have made significant advances in developing strategies to generate effective immune responses against ovarian cancer. We are currently validating these approaches in clinical trials. We anticipate that this immunotherapy approach will ultimately be most beneficial for patients who have completed their standard treatments for ovarian cancer to minimize the risk of relapse.

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