Understanding Primary Peritoneal Cancer

A Woman’s Guide

Gynecologic Cancer Foundation
The Gynecologic Cancer Foundation (GCF) is a 501(c) 3 not-for-profit organization whose mission is to ensure public awareness of gynecologic cancer prevention, early diagnosis and proper treatment. In addition, the Foundation supports research and training related to gynecologic cancers. GCF advances this mission by increasing public and private funds that aid in the development and implementation of programs to meet these goals.
## CONTENTS

<table>
<thead>
<tr>
<th>Topic</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>INTRODUCTION</td>
<td>2</td>
</tr>
<tr>
<td>PRIMARY PERITONEAL CANCER: AN OVERVIEW</td>
<td>3</td>
</tr>
<tr>
<td>RISK FACTORS</td>
<td>4</td>
</tr>
<tr>
<td>MEDICAL EVALUATION</td>
<td>5</td>
</tr>
<tr>
<td>WORKING WITH YOUR TREATMENT TEAM</td>
<td>8</td>
</tr>
<tr>
<td>TREATMENT</td>
<td>11</td>
</tr>
<tr>
<td>ONCE YOU HAVE BEEN TREATED, THEN WHAT?</td>
<td>15</td>
</tr>
<tr>
<td>RECURRENT DISEASE</td>
<td>16</td>
</tr>
<tr>
<td>LIVING WITH CANCER THERAPY</td>
<td>17</td>
</tr>
<tr>
<td>TIPS FOR COPING</td>
<td>20</td>
</tr>
<tr>
<td>RESOURCES FOR MORE INFORMATION</td>
<td>23</td>
</tr>
</tbody>
</table>
Introduction

You and your family have learned of a diagnosis of primary peritoneal cancer (PPC). The amount of information you receive at the time of diagnosis can feel overwhelming. All at once, you may feel there are questions to be answered, decisions to be made, and so much information to be understood.

A team of healthcare professionals will work with you throughout your treatment process. Each of them has an important job, but the most vital member of the team is you. To play an active role during your treatment, it helps to learn as much about primary peritoneal cancer as possible.

This booklet will take you through the basics of what you need to know about primary peritoneal cancer, or PPC. It will introduce you to the people who may be part of your treatment team. It will identify the different types of treatment for PPC. It may also help prepare you to talk with your treatment team and to feel more confident about your treatment plan.
Cancer occurs when cells in an area of the body grow in an abnormal way. Primary peritoneal cancer is a relatively rare cancer that develops most commonly in women. PPC is a close relative of epithelial ovarian cancer, which is the most common type of malignancy that affects the ovaries. The cause of primary peritoneal cancer is unknown.

It is important for women to know that it is possible to have primary peritoneal cancer even if their ovaries have been removed.

The abdominal cavity, as well as the surface of all the organs in the abdomen, are covered in a cellophane-like, glistening, moist sheet of tissue called the peritoneum. It not only protects the abdominal organs, it also supports and prevents them from sticking to each other and allows them to move smoothly within the abdomen.

The cells of the peritoneal lining develop from the same type of cell that lines the surface of the ovary. Certain cells in the peritoneum can undergo transformation into cancerous cells, and when this occurs, the result is primary peritoneal cancer. It can occur anywhere in the abdominal cavity and affect the surface of any organ contained within it.
RISK FACTORS

Similar to most cancers, the aging process is associated with a higher chance of developing peritoneal cancer. Therefore, the major risk factor for primary peritoneal cancer is advancing age. There are certain families in whom ovarian and breast cancers are common. Many of these families carry a genetic mutation in a gene, which leads to a dramatically increased risk of developing ovarian cancer and PPC, as well as fallopian tube cancer and certain types of uterine cancer. However, only 10% of ovarian and primary peritoneal cancers are genetically linked.

SYMPTOMS

Unfortunately, because of the vague nature of its symptoms, PPC is usually diagnosed in advanced stages of disease, when achieving a cure is difficult. The symptoms of PPC are more commonly gastrointestinal rather than gynecologic in nature, and include abdominal bloating, changes in bowel habits and an early feeling of fullness after eating. When bloating is severe, nausea and vomiting may result. Occasionally, patients with PPC present with a blockage of the intestines related to tumor on or next to the bowels. Normal vaginal bleeding is infrequently seen in patients with PPC.
Primary peritoneal cancer is usually diagnosed when a woman sees her doctor complaining of abdominal swelling and bloating. As described above, the symptoms of PPC are more commonly gastrointestinal than gynecologic in nature. These symptoms are related to the accumulation of fluid, also known as ascites, that occurs with PPC. Gastrointestinal symptoms also occur because seedlings of tumor often line the peritoneal surface (the outer lining) of the intestines, a process called carcinomatosis. The omentum, an apron of fatty tissue that hangs down from the colon and stomach, often contains bulky tumor, described as omental caking. Although these findings can be detected on a physical exam, they frequently are subtle and difficult to detect. When a woman is found to have fluid in the abdomen (ascites), the usual first step toward a diagnosis is a CT scan. This is a special type of x-ray test that allows doctors to assess the entire abdomen and pelvis. Omental caking and ascites, as well as other tumor growths, are commonly seen, and point toward the diagnosis of PPC or ovarian cancer. Other cancers can cause these findings, thus, further tests are needed and are usually focused around ruling out other more common cancers, such as colon and breast cancer.

Frequently, the evaluation of ascites begins with a procedure known as a paracentesis, whereby fluid is removed from the abdomen using a needle. The fluid is examined under the microscope, looking for the presence of cancerous cells. Unfortunately, this procedure is not without risks as the process of performing a paracentesis can actually “seed” the abdominal wall with cancer cells. Therefore, it is important to seek the advice of a gynecologic oncologist when considering this procedure as it may not be necessary given that most patients with these findings will undergo surgery regardless of the results. However, it may be helpful in the patient that is either not a surgical candidate or in one suspected of having ascites for reasons other than cancer, such as liver or heart disease.
Another test that is commonly performed when PPC is suspected is the CA125 blood test. CA125 is a chemical that is made by tumor cells and is usually elevated in patients with PPC. Unfortunately, it can also be elevated in a variety of benign conditions, as well as other cancers, and thus an elevated CA125 blood test does not mean the patient has PPC. For more information about CA125, please visit the Gynecologic Cancer Foundation’s Web site (www.thegcf.org) for a brochure entitled “Understanding CA125 Levels.”

The actual diagnosis of PPC is often not completely certain until a woman undergoes surgery. This is because the clinical presentation of the disease is the same as that of epithelial ovarian cancer. Furthermore, PPC and epithelial ovarian cancer also appear identical under the microscope. It is, therefore, the pattern of tumor distribution in the abdominal cavity that often indicates primary peritoneal rather than ovarian cancer. Patients with PPC are usually found to have normal ovaries, or only superficial involvement of the ovaries at the time of pre-surgical imaging tests or at time of surgery. However, the diagnosis can occasionally remain uncertain even following surgery. It is important to understand that PPC can occur in women whose ovaries have already been removed.

**SURGICAL STAGING**

Surgical staging of cancers is performed in order to fully assess the extent of disease. This allows for decisions to be made regarding additional therapy, which is usually in the form of chemotherapy. Surgical staging generally involves removal of all visible disease, as well as removal of the ovaries, fallopian tubes and uterus, it can also include removal of the omentum, lymph nodes and other organs depending on the surgical findings. It is imperative that this surgery be performed by a gynecologic oncologist. These specialist are most familiar with the treatment of this
cancer, thus offering patients the best chance of survival. There is no formal agreed-upon staging system for primary peritoneal cancer. Because it is so similar to ovarian cancer with respect to treatment, it is staged in a similar fashion. Tumor state is typically assigned using guidelines established for ovarian cancer. Stages I through IV describe how far the tumor has spread. Nearly all patients diagnosed will have Stage III or higher because warning signs are typically few until the cancer is widespread.

Patients with primary peritoneal cancer may have fluid around the lungs, known as a pleural effusion. If an effusion is present, some fluid may be removed in order to look for tumor cells. If tumor cells are found in this fluid, the patient has Stage IV disease.
During your treatment, you will come in contact with many healthcare professionals. These people make up your treatment team. They will work with each other and with you to provide the special care you need. Your treatment team may include some of the healthcare professionals listed below.

Ideally, your treatment will be provided and managed by a gynecologic oncologist. Gynecologic oncologists are obstetrician-gynecologists who have an additional 3-4 years of special training in the comprehensive surgical care and medical treatment of female reproductive cancers including primary peritoneal cancer. A gynecologic oncologist can manage your care from diagnosis to completion of treatment. Women with primary peritoneal cancer who have their cancer surgery done by a gynecologic oncologist have higher cure rates than women who have surgery done by any other type of physician. The better survival is related to the fact that gynecologic oncologists are more likely to remove more of the cancer at the time of surgery and more accurately determine the stage of the cancer.
You also may be treated by a:

➤ **Medical oncologist** who specializes in using drug therapy (chemotherapy) to treat cancer.

➤ **Radiation oncologist** who specializes in using radiation therapy to treat cancer.

➤ **Oncology nurse** who specializes in cancer care. An oncology nurse can work with you on every aspect of your care, from helping you understand your diagnosis and treatment to providing emotional and social support.

➤ **Social worker** who is professionally trained in counseling and practical assistance, community support programs, home care, transportation, medical assistance, insurance and entitlement programs. They are very helpful advocates, especially when you are first diagnosed and unsure what to do next.

➤ **Nutritionist or registered dieticians** who are expert in helping you either maintain or initiate healthy eating habits. This is important in the recovery process. These professionals can help you overcome potential side effects of treatment such as poor appetite, nausea or mouth sores. It is important to note that natural remedies and supplements should be taken only under the supervision of a naturopathic physician in consultation with your gynecologic oncologist.
Talking with your treatment team

You deserve expert advice and treatment from your treatment team. Be sure to talk openly about your concerns with the members of your treatment team. Let them know what is important to you. If it is hard for you to speak for yourself, these tips may help:

➤ Make a list of questions before your visit. Ask the most important questions first.

➤ Take notes or ask if you can tape record your medical office visits and phone conversations.

➤ If you don’t understand something, ask the treatment team member to explain it again in a different way.

➤ If possible, bring another person with you when you meet with members of your treatment team to discuss test results and treatment options.
**TREATMENT**

Primary peritoneal cancer is treated in the same way ovarian cancer is treated. It is most often treated with surgery and chemotherapy. Only rarely is radiation therapy used. Your specific treatment plan will depend on several factors, including:

- The stage and grade of your cancer
- The size and location of your cancer; and
- Your age and general health

All treatments for primary peritoneal cancer have side effects. Most side effects can be managed or avoided. Treatments may affect unexpected parts of your life, including your function at work, home, intimate relationships, and deeply personal thoughts and feelings.

Before beginning treatment, it is important to learn about the possible side effects and talk with your treatment team members about your feelings or concerns. They can prepare you for what to expect and tell you which side effects should be reported to them immediately. They can also help you find ways to manage the side effects you experience.

**SURGERY**

Surgery is usually the first step in treating primary peritoneal cancer and it should be performed by a gynecologic oncologist. The goal of the surgery is the removal of all visible disease because this approach has been shown to improve survival. This process is known as tumor debulking. When all visible disease is removed, or if only small tumor implants (less than 1 cm in diameter) remain, the patient is considered optimally debulked. Occasionally, the location of tumor within the abdomen or the condition of the patient does not allow for optimal debulking surgery to
be performed. In this situation, chemotherapy may be given first and the patient might have surgery at a later time. Most surgery is performed using a procedure called a laparotomy during which the surgeon makes a long cut in the wall of the abdomen. If PPC is found, the gynecologic oncologist performs the following procedures:

- **Salpingo-ooophorectomy:** both ovaries and fallopian tubes are removed.
- **Hysterectomy:** the uterus is removed usually with the attached cervix.
- **Omentectomy:** the *omentum*, a fatty pad of tissue that covers the intestines, is removed.

Occasionally, some of the nearby lymph nodes will be removed. Depending on the surgical findings, more extensive surgery, including removal of portions of the small or large intestine and removal of tumor from the liver, diaphragm and pelvis, may be performed. Removal of as much tumor as possible is one of the most important factors affecting cure rates.

**Side effects of surgery**

Some discomfort is common after surgery. It often can be controlled with medicine. Tell your treatment team if you are experiencing pain. Other possible side effects are:

- Nausea and vomiting
- Infection, fever
- Wound problem
- Fullness due to fluid in the abdomen
- Shortness of breath due to fluid around the lungs
- Anemia
➤ Swelling caused by lymphedema, usually in the legs or arms
➤ Blood clots
➤ Difficulty urinating or constipation

Talk with your doctor if you are concerned about any of the problems listed.

**CHEMOTHERAPY**

Chemotherapy is the use of drugs to kill cancer cells. It can be given intravenously (injected into a vein) or, more recently, intraperitoneal administration has become popular because it is associated with a longer survival in patients with a very similar cancer, ovarian cancer. Intraperitoneal chemotherapy involves the administration of medicines directly into the abdomen through a catheter which is placed under the skin at the time of initial surgery, or shortly thereafter. Unfortunately, it has more immediate side effects than intravenous chemotherapy and therefore some patients prefer the more traditional intravenous administration. Intraperitoneal treatment is only given if optimal debulking surgery has been achieved. Either treatment may be administered in the doctor’s office, outpatient treatment areas of the hospital or occasionally, as an inpatient.

Traditionally, intravenous chemotherapy is given on an every three-week schedule as an outpatient. Each treatment of chemotherapy is known as a cycle and initial treatment usually consists of six cycles. Intraperitoneal chemotherapy is also given on an every three-week schedule for six cycles. But, each cycle is a little more involved as the patient might receive treatments on several days of the 21 day cycle compared to receiving treatments on only day 1 of the cycle if given intravenously.

The most commonly used chemotherapy medicines for PPC are the same as those used for ovarian cancer. These include one of the platinum-based medicines, Cisplatin or Carboplatin, as well as ataxane (Paclitaxel or Taxotere) in combination.
Side effects of chemotherapy

Each person responds to chemotherapy differently. Some people may have very few side effects while others experience several. Most side effects are temporary. They include:

➤ Nausea
➤ Loss of appetite
➤ Mouth sores
➤ Increased chance of infection
➤ Bleeding or bruising easily
➤ Vomiting
➤ Hair loss
➤ Fatigue
➤ Constipation
➤ Diarrhea

RADIATION

Radiation therapy may be utilized for treatment of isolated small areas of disease that has returned after initial therapy. It is rarely used as a first therapy for PPC.
Once you have been treated, then what?

After initial treatment is completed, patients with primary peritoneal cancer are followed closely with visits every 2 to 4 months for the first 3 years and then every 6 months. At each visit they have a physical exam, including a pelvic exam, CA125 testing, and, depending on the patient and her situation, imaging tests, such as CT scans, X-rays, MRIs or PET scans, may be performed. Because PPC has a tendency to recur with time, patients often require more than one round of chemotherapy and may also need additional surgical procedures.
Recurrences are common in patients with PPC because most patients with PPC are diagnosed when they already have advanced stages of disease. The majority of patients will initially go into remission, but the disease commonly returns months to years later when the CA 125 levels begin to rise or new masses are found on physical exam or imaging studies. Unfortunately, the prognosis for this cancer is not favorable once it recurs, but a longer remission before recurrence is associated with a better chance for a second, third and even fourth remission.

There are several treatment options for patients who recur, depending on the location of recurrence, time since the initial therapy and the patient’s overall performance status. These options include repeat surgery, re-treatment with the same chemotherapy that was given initially or a different type of agent. They can also consider radiation therapy for selected cases. Each recurrence is different, so their treatment must be individualized based on a variety of factors including those listed above. It is also important to investigate whether there is a clinical trial that is appropriate for the patient. Unfortunately, once a recurrence is diagnosed, one must re-focus the goals of treatment to help prolong quality of life rather than a cure.
The experience of being diagnosed with primary peritoneal cancer and undergoing cancer treatment may change the way you feel about your body, and will affect your life in many ways. You may experience many or relatively few side effects. Being aware of the possible treatment effects may help you anticipate them and plan ways to cope.

Fatigue
Regardless of the treatment prescribed, you are likely to experience fatigue and there will be times when you do not feel well enough to take care of tasks at home. You probably will need to rely on family and friends to help with some of the things you usually do. You may want to consider hiring someone for help with chores until you feel well enough to manage again. If you know that you will not have support at home, talk frankly with your healthcare team as early as possible so that alternatives can be explored. Since a nourishing diet is important, be sure to ask for help, if needed, in maintaining healthy meal and snack choices in your home.

Be sure that your blood count is checked to rule out anemia as a treatable cause of fatigue. There are also medications for the relief of fatigue.

Work life
You will probably need to be away from work quite a bit during the first month or two of your treatment. Talk with your supervisors at work and with your healthcare team to set up a realistic plan for work absences and return to work. Remember to tell your work supervisor that any plan must be flexible because your needs may change as treatment progresses. The Family Medical Leave Act (FMLA) offers certain protections for workers and family members who must be away from work for health reasons.
Facing the world
The effects of cancer and your cancer treatment may alter your appearance. You may appear fatigued, pale, slow moving, and you may have to face temporary hair loss. You may feel self-conscious because of these changes. It might help to imagine how you would feel if you saw a friend or sister looking as you do. Remember that many people are loving you rather than judging you as they notice these changes.

Family, friendships and fun
No matter what type of treatment you have for your primary peritoneal cancer, you may experience side effects that could affect how you feel about joining in social events with friends and family. Talk to your healthcare team if special events are coming up, such as a wedding or graduation. The timing of your treatments may be able to be adjusted so that you feel as well as possible for these special days. Don’t hesitate to plan activities that you enjoy. You may have to cancel on occasion or leave a little early, but the good times will help you to find strength for the hard days.

It is often difficult for young children to understand what you are going through. Counselors are available to help you answer questions and to help your children cope. It is also a good idea to ask family and friends to help you keep your children’s normal routine.

Driving
For women who drive, driving is an almost indispensable part of adult life. You should not drive if you are taking medications that cause drowsiness, such as narcotic pain relievers and some nausea medications. Most women can start driving again within a few weeks of surgery and usually women can drive most days during chemotherapy and radiation therapy. Be sure to ask your healthcare team about driving.
Sexuality

Some treatments for primary peritoneal cancer can cause side effects that may change the way you feel about your body, or make it difficult to enjoy intimate or sexual relationships. Which side effects you experience depend on your treatment course. You may experience some or none at all. Being aware of the possible side effects may help you anticipate them and learn ways to cope with them.

Possible side effects include:

➤ **Hair loss.** A common side effect of chemotherapy, hair loss is usually temporary. Still, it can be difficult to accept. If you experience hair loss, you may choose to wear flattering wigs, scarves or other headwear.

➤ **Vaginal changes.** Some forms of treatment, such as hysterectomy and radiation therapy, may cause dryness, shortening and narrowing of the vagina. These changes can make sexual activity uncomfortable. Using an over-the-counter vaginal lubricant may help you feel more comfortable. Your treatment team may also recommend a vaginal dilator.

➤ **Reduced sexual desire.** The stress and fatigue you may experience during cancer treatment may cause you to lose interest in sex for a period of time.
TIPS FOR COPING

Talk with your treatment team. Your treatment team members can provide advice based on your individual situation, so it is very important that you talk honestly with them. You may want to ask:

➤ How will my treatment affect my sexuality?
➤ Will these effects be temporary or permanent?
➤ Are there other treatment options that might lessen these effects?
➤ Do you have suggestions about how I can deal with the effects of treatment on my sexuality?

Communicate with your partner. Having cancer can strain both partners in a relationship. Talking about the sexual and emotional effects cancer has on your relationship can be difficult. But you may find it easier to work through the challenges if you talk through them together. Be prepared to share your own feelings and to listen to what your partner has to say.

Shift your focus to intimacy. Sexual intercourse is only one part of intimacy. You may find that touching, kissing and cuddling are equally fulfilling.

Be patient with yourself. Understand that a return to a sexual relationship may take time. Your treatment team can tell you if and how long you should wait to have sex after treatment. It may be awhile before you feel emotionally ready. Give yourself the time you need.

Keep an open mind. Having an open mind and a sense of humor about ways to improve your sexuality may help you and your partner find what works best for you. For more information, please visit the Gynecologic Cancer Foundation’s Web site (www.thegcf.org) for a brochure entitled “Renewing Intimacy and Sexuality after Gynecologic Cancer.”
Seek support. There are many resources available to help you deal with any sexual or emotional issues you may have as result of cancer and its treatment. Specially trained counselors can help you deal with the impact of cancer on your life. Support groups are another good resource. People who are facing a situation similar to yours can come together to share their experiences and give one another advice and emotional support. To find support services in your area, talk with a member of your treatment team, or contact the resources listed on pages 23 and 24 of this brochure.

Exercise
During treatment you may find that even the stairs to your bedroom are a challenge even if you have worked hard during your adult life to keep fit. It's discouraging, but normal, to have to reduce or interrupt your fitness routine. If you’ve had surgery, ask your doctor for specific guidelines about exercise. During chemotherapy or radiation, adjust your exercise according to how you feel.

You should avoid overexerting or dehydrating yourself. Over the weeks and months after you finish cancer treatment, you can build back toward your previous level of fitness.

Hopeful messages
As you go through cancer treatment, be patient with yourself. Understand that a return to your full and wonderful life will take time. Your treatment team can guide you through the difficulties that you will face if they know what is troubling you. Talk openly about the things that bother you. Give yourself the time you need.

Advance Medical Directives can be a helpful tool for clarifying your medical care wishes. We encourage both patients and family members to complete one. Your healthcare team is available for guidance on this matter.
Nurture hope. It’s up to you to take charge of your reaction even as you face the unknown of cancer. Hope helps you see the positive aspects of life.

If you have inner spiritual beliefs, reach out to your religious community to give you additional support to face each day and LIVE.

Seek support. There are many resources available to help you deal with the physical, sexual or emotional issues you may have as result of cancer and its treatment. Specially trained counselors can help you deal with the impact of cancer on your life. Support groups are another good resource. People who are facing a situation similar to yours can come together to share their experiences, and give one another advice and emotional support. To find support services in your area, talk with a member of your treatment team, or contact the resources listed on pages 23 and 24 of this brochure. Remember you are surrounded by a devoted healthcare team, so let us be at your side.
RESOURCES FOR MORE INFORMATION

American Cancer Society (ACS)
800.ACS.2345 • www.cancer.org

American College of Obstetricians and Gynecologists (ACOG)
202.863.2518 • www.acog.org • resources@acog.org

CancerCare
800.813.HOPE • www.cancercare.org • info@cancercare.org

Centers for Disease Control and Prevention (CDC)
800.CDC.INFO (800.232.4636) • www.cdc.gov/cancer • cancerinfo@cdc.gov

CONVERSATIONS!
The International Ovarian Cancer Connection
806.355.2565 • www.ovarian-news.org • ovcanews@aol.com

EyesOnThePrize.org Support for Gynecologic Cancer
www.eyesontheprize.org • info@eyesontheprize.org

Force: Facing Our Risk of Cancer Empowered
866.288.RISK • www.facingourrisk.org • info@facingourrisk.org

Gilda’s Club Worldwide
888.GILDA.4.U • Fax: 917.305.0549 • www.GildasClub.org • info@gildasclub.org

Gilda Radner Familial Ovarian Cancer Registry
800.OVARIAN • 716.845.4503 • www.ovariancancer.com • gradner@roswellpark.org

Gynecologic Oncology Group
800.225.3053 • www.gog.org • kness@gog.org

Gynecologic Cancer Foundation (GCF)
312.578.1439 • 800.444.4441 (Information Hotline)
www.thegcf.org or www.wcn.org • info@thegcf.org

In My Sister’s Care (IMSC)
202.607.1883 • 212.802.7686 • www.inmysisterscare.org • info@inmysisterscare.org
TAKE ACTION — TELL A FRIEND

Know your risk

➤ Advancing age

➤ Family history of ovarian, fallopian tube or premenopausal breast cancer

➤ Log-on to the Women’s Cancer Network (www.wcn.org) to take a confidential risk assessment

Know the symptoms

➤ Abdominal bloating

➤ Changes in bowel habits

➤ Early feeling of fullness after eating

➤ If primary peritoneal cancer is suspected, consult a gynecologic oncologist

Women treated by gynecologic oncologists are more likely to get appropriate surgery and have higher cure rates.

To find a gynecologic oncologist in your area, contact the Gynecologic Cancer Foundation’s Information Hotline at 1.800.444.4441 or log onto the Women’s Cancer Network (www.wcn.org) and enter your zip code in the “Find-A-Doctor” section.