What to Do Next:

When the Diagnosis Is Mucinous Ovarian Cancer

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WHEN THE DIAGNOSIS IS MUCINOUS OVARIAN CANCER

The team at the Mucinous Ovarian Cancer Coalition (MOCC) hears this question often: *what guidance would we share if someone we loved received the diagnosis of mucinous ovarian cancer?* It's a tough one for us to answer.

If you aren't familiar with how MOCC came to be, we encourage you to visit the About section of our website (*hope4moc.org*) to read ***Rhonda's Story***.

Rhonda was our late cofounder. Every step of her journey, beginning with a missed diagnosis, was a tale of how this disease should **not** be handled. To prevent other women from suffering a similar experience, we asked our past Physician Advisory Committee Chair, the late Dr. Michael Frumovitz of the MD Anderson Cancer Center, for advice.

THE NEXT STEPS: HOW TO NAVIGATE A DIAGNOSIS OF MUCINOUS OVARIAN CANCER

Work with a Gynecologic Oncologist

The first thing I would make sure of is that my loved one is under the care of a gynecologic oncologist. Gynecologic oncologists are trained to take care of women with cancers of the ovary, uterus, cervix, vagina, and vulva. For a rare tumor like mucinous ovarian cancer, a gynecologic oncologist will have the most experience and expertise in deciding the best options for care.

When a woman is found to have mucinous cancer on the ovary, the first important step is to determine whether the malignancy started on the ovary (primary mucinous ovarian cancer) or spread to the ovary from another organ (metastatic disease). Over 50% of the time, the disease started somewhere else, such as in the pancreas, small bowel, appendix, or colon.

Determining Where the Disease Started

A general "rule of thumb" is that if both ovaries have tumors that are less than 10 cm in size, then it is likely metastatic disease. If a tumor is found on only one ovary and is greater than 10 cm in size, it is likely a primary mucinous ovarian cancer.

I would want a well-trained, experienced pathologist evaluating the ovary to determine site of origin because treatment for primary ovarian cancer is frequently different than it is for a gastrointestinal cancer. Smaller hospitals may not have such a pathologist on staff, so in those cases I would strongly recommend having the pathology specimens sent to an expert pathologist at another institution.

If the pathologist believes that the patient indeed has a primary mucinous ovarian cancer, evaluation of the gastrointestinal system with a colonoscopy and an upper endoscopy (a camera put down the throat to look at the stomach and small bowel) should be considered to confirm the diagnosis. The patient should also have a CT scan of the chest, abdomen, and pelvis to look for other sites of disease.

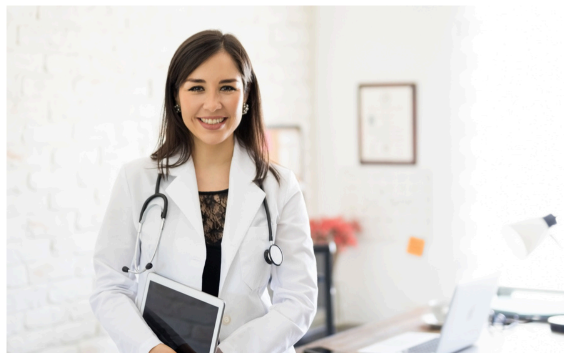
Sometimes patients ask if they should have a PET scan instead, but a PET scan is not any better than a CT scan in detecting this type of cancer.

Is Another Surgery Necessary?

If the woman had unknown disease at the time of her surgery and had just one ovary removed, a decision as to whether another surgery is necessary will be discussed. Many factors go into that decision, including the patient's age, her desire for future fertility, CT scan findings, certain pathologic findings, and the patient's wishes.

This decision can be very complex and reiterates the importance of having a gynecologic oncologist as part of the care team. Over 80% of the time this cancer will be limited to just one ovary, so if the patient is young and the remaining ovary looks normal, I would strongly recommend not removing the other, normal-appearing ovary or the uterus. A number of my patients have gotten pregnant and had healthy children after a diagnosis of primary mucinous ovarian cancer.

Is Chemotherapy the Next Step?



Another important decision is whether a patient needs chemotherapy after surgery, and what type of chemotherapy to give. If disease is found on other organs besides the ovary, then chemotherapy is definitely recommended. If disease is found on just one ovary, then the gynecologic oncologist may recommend chemotherapy based on certain pathologic features of the tumor.

Currently there are two schools of thought about what type of chemotherapy to give a patient with primary mucinous ovarian cancer—and both options are reasonable.

> Some gynecologic oncologists recommend the drugs typically given for other types of ovarian cancer, which are usually carboplatin and paclitaxel (Taxol).

> Others believe that because this tumor looks and behaves more like a gastrointestinal cancer, it should be treated with drugs like 5-FU, oxaliplatin, and irinotecan that are typically used for that type of cancer.

I would want my loved one to get the types of chemotherapies used for gastrointestinal cancer as I believe these work better than the standard ovarian cancer treatments. To be clear, however, giving either type of chemotherapy is currently acceptable.

Follow Up After Chemotherapy

Once my loved one has completed therapy, we would watch her "like a hawk." So, what does that look like? She would be seen every 3 months for the first year after completing therapy, then every 4 months in the second year, and then every 6 months for three years after that. At those appointments we would talk about any symptoms she may be having (like pain, nausea, or bloating) and would do a physical exam including a pelvic exam.

We would also get laboratory tests for tumor markers. These tests look for certain proteins in the blood that may be elevated in different types of cancers. The markers I typically follow are CA125, CEA, and CA19-9. If any of these markers is elevated, I would be concerned that the cancer had recurred and would perform further evaluation, typically with a CT scan.

If, however, the exam is negative, the patient has no symptoms, and the markers are normal, we would not get a CT scan. In other words, a CT scan is not warranted at every visit, but only if we suspect a recurrence. However, if the patient still has a remaining ovary, I would recommend getting a pelvic ultrasound at each of those appointments.

When Mucinous Ovarian Cancer Recurs

If my loved one were to unfortunately have a recurrence of her disease, I would look for a clinical trial for her. At MD Anderson we opened a clinical trial that includes repeat surgery to remove the tumor and HIPEC (hyperthermic intraperitoneal chemotherapy), which is heated chemotherapy given directly into the abdomen to bathe any remaining tumor deposits.

Other trials may use newer drugs that target the mutations in the tumor cells that lead to the cancer formation. For example, a mutation in the KRAS gene is present in over 50% of primary mucinous ovarian cancer tumor specimens.

There are multiple drugs that target the KRAS mutation, many of them available through clinical trials. These mutations are detected with a tumor test called molecular testing.

It is important that all patients with recurrent disease get this testing done on the tissue from either their original surgery or a biopsy. Some centers do this testing themselves, while others outsource it to biotech companies. The most popular companies that perform this testing are Foundation Medicine and Caris.

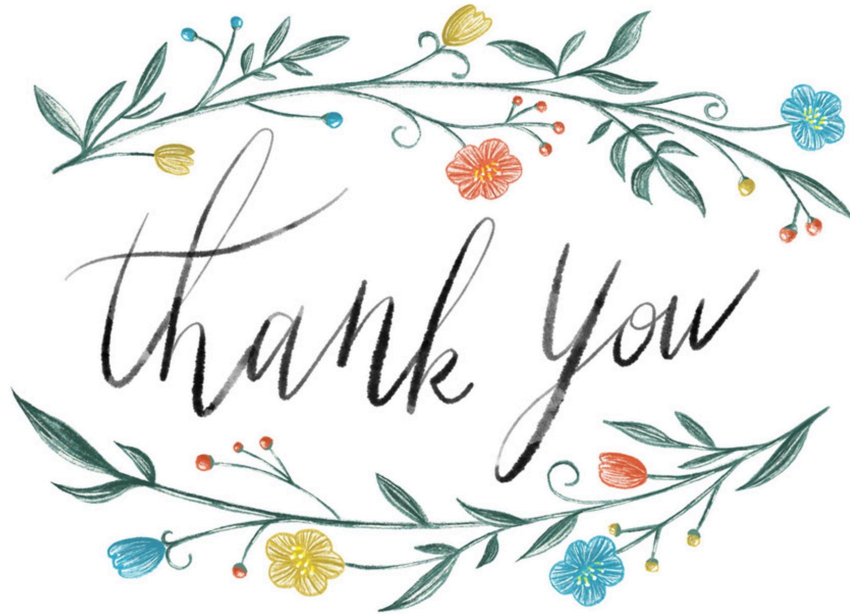
Quality of Life Is Important

Throughout the entire journey, it would be essential for my loved one to always consider how any treatment will affect her quality of life. If disease were to recur, it is unlikely that it will be curable, so all decisions about more treatment need to consider how much time the treatment will add to her life and how the side effects may impact her quality of life.

It would also be wise to get a palliative or supportive care team involved early in the process, as this has been shown to not only improve quality of life in patients undergoing treatment for recurrence but also may increase survival time. I would recommend getting a palliative care team involved right at the time of recurrence, rather than waiting until the patient is out of treatment options or has significant symptoms.

The Bottom Line

In summary, most women with primary mucinous ovarian cancer will have early-stage disease (limited to one ovary) and hence will have a high likelihood of being cured. For a woman with metastatic or recurrent disease, the chance of cure is small, but additional treatment may extend her life. However, those treatments should always be considered in light of their side effects and how they will affect her quality of life.



The Mucinous Ovarian Cancer Coalition (MOCC) continues to remember and appreciate the work of our late Physician Advisory Chair, Dr. Michael Frumovitz.

He donated his time and expertise to create this guide. We know it will help women who are newly diagnosed with this disease.

Visit Hope4MOC.org to learn more