



# Malignant Ovarian Germ Cell Tumors

## WHAT SHOULD I KNOW?



### EPIDEMIOLOGY

Ovarian germ cell tumors (OGCTs) comprise only 5% of all >21,000 ovarian cancers diagnosed annually in the United States. They tend to affect children and women younger than 40 years and arise from the primordial germ cells of the ovary, which develop into eggs. Many secrete proteins that can be measured in the bloodstream known as ‘tumor markers,’ such as LDH (lactate dehydrogenase), HCG (human chorionic gonadotropin), AFP (alpha fetoprotein), estrogens (i.e., estradiol, E2) and androgens (A). There are several subtypes of malignant OGCTs:

SUBTYPE	TUMOR MARKER				
	LDH	HCG	AFP	E2	A
dysgerminomas	+	±	-	±	-
yolk sac tumors	+	-	+	-	-
embryonal carcinomas	±	+	±	±	-
polyembryoma	-	+	±	-	-
non-gestational choriocarcinoma	±	+	-	-	-
immature teratoma*	±	-	±	-	±
mixed OGCT	±	±	±	±	±
gonadoblastoma†	-	-	-	±	±

\* mature teratomas (dermoid cysts) are non-cancerous but in approximately 1% of cases portions of the tumor can degenerate into a cancer

† gonadoblastomas are a non-invasive tumor with malignant potential

### SIGNS & SYMPTOMS

At least 85% of patients present with abdominal pain and an abdominal mass. If the mass ruptures (10%), the pain can have a rapid onset. These tumors often grow quickly and involve both ovaries in 10% of instances. Tumors that secrete HCG can cause children to experience precocious (early) puberty and may produce abnormal vaginal bleeding in older women.

### SCREENING

There are no good screening tests for this condition. Patients with gonadal dysgenesis (harboring a Y chromosome) or Turner’s syndrome are at risk of developing a gonadoblastoma and could benefit from prophylactic oophorectomy (removal of the ovary).

### TREATMENT

**After Surgery:** Unlike many epithelial ovarian cancers, fertility-sparing surgery can often be performed safely and is recommended for any individual who desires childbearing. This could include the removal of one ovary and fallopian tube, with biopsies of lymph nodes, the omentum, and lining of the abdomen. Surgery establishes the diagnosis, allows the tumor to be staged (extent of spread at the time of diagnosis), and is also the first step in treatment. The surgery may be performed using either laparoscopic (several small incisions) or open (one large incision) techniques.

**Chemotherapy:** Surgery alone can be sufficient treatment for stage I dysgerminoma and stage I grade I immature teratomas. All other patients generally benefit from chemotherapy, which often includes 3-4 cycles of bleomycin + etoposide + cisplatin

OR

carboplatin + etoposide

Children and young adults who receive etoposide may be at risk for developing a second cancer later in life (secondary malignancy) such as leukemia. More than 80% of patients treated with these regimens resume menses, in which case, pregnancy outcomes are generally good. Additional chemotherapies may be effective for recurrence.

**Immunotherapy:** If a tumor displays certain characteristics (microsatellite instability, high mutational burden) it may respond to immunotherapy, such as pembrolizumab.

### FOLLOW UP

Upon remission, patients with germ cell tumors should undergo reassessment (clinical exam and biomarkers) every 3 months for the first 2 years then every 6 months until 5 years after completing of treatment. After 5 years, patients can return to annual exams. Stage is the most important factor to determine survival outcomes, which approach at 5 years 100% for stage disease and 70% for stage IV disease.

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### QUESTIONS YOU SHOULD ASK YOUR CARE TEAM ABOUT YOUR TREATMENT PLAN and FOLLOW-UP CARE

Does my cancer have a tumor marker?

What was my stage at diagnosis?

Should I be evaluated by fertility specialist?

Are there any clinical trials available for my disease?

If you are not already being treated by a gynecologic oncologist, consider seeking a second opinion.

Things to consider when getting a second opinion:

- Pathology re-review...
- Tumor board...

#### SOURCES & MORE INFORMATION:

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