



WORLD  
**OVARIAN**  
CANCER  
COALITION



*Empowering the  
global ovarian cancer  
community through  
knowledge, collaboration,  
and action.*

## RARE OVARIAN CANCER

This briefing is intended to provide a high-level overview of rarer forms of ovarian cancer, the challenges that face those impacted by these diseases, and the actions needed to address unmet need in this area.

This briefing is a follow-up to a panel discussion held by the World Ovarian Cancer Coalition in 2022 as part of our Global Annual Partner Meeting.

April 2023

**NO  
WOMAN  
LEFT  
BEHIND**

# RARE OVARIAN CANCER

## TYPES OF OVARIAN CANCER

The World Health Organization (WHO) lists many types of ovarian cancer and categorizes the disease into three groups:<sup>1 2:</sup>

- **Epithelial tumours** - the most common form of ovarian cancer and occurs primarily in adults
- **Germ cell tumours** - occurs primarily in children and teens and is rare compared to epithelial ovarian tumours
- **Stromal tumours** – rare compared to epithelial tumours

About 85-90% of malignant ovarian cancers are epithelial ovarian carcinomas<sup>3</sup> (cancerous epithelial tumours are called carcinomas). There are many sub-types of cancerous epithelial tumours, the serous type is by far the most common and can include high-grade and low-grade tumours. The approximate 10% of remaining ovarian cancers develop from germ cells or granula-theca cells<sup>4</sup>.

## TYPES OF RARE OVARIAN CANCER

About 10-15% of ovarian tumours are considered rare<sup>5</sup>. Patients with rare ovarian cancers frequently feel isolated because of the difficulties in finding a specialist with expertise in their cancer. The survival rates of rare types of ovarian cancer depends on the specific sub-type. For example, low grade serous ovarian cancer, which accounts for approximately 10% of epithelial ovarian cancers, is hard to treat and has an average survival rate of 9 years<sup>6</sup>.

### *Rare Types of Epithelial Ovarian Cancer*

As mentioned, by far the the most common type of epithelial ovarian cancer is high-grade

***“We are at a disadvantage as a whole community because of the underfunding of rare ovarian cancers... just 6% of ovarian cancer funding went to rare ovarian cancers [UK, 2007-2019], significantly less than you would expect.”***

***Jane Ludemann  
Founder  
Cure Our Ovarian Cancer***

serous carcinoma, which accounts for 70% of diagnoses<sup>7</sup>. The remaining 30% are rare and fall mainly into the sub-types of mucinous carcinoma, clear cell carcinoma, endometrioid or low-grade serous carcinoma<sup>8</sup>, although there are others<sup>9</sup>. These main sub-types have similarities and differences, one major difference among them is that mucinous carcinoma, clear cell carcinoma, and, to some degree, endometrioid carcinoma typically present in the early stages - either stage II or III. In contrast, low-grade serous carcinoma is more likely to present in stage III or IV<sup>10</sup>.

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**“We don’t have any evidence-based treatment option for mucinous, so we’re not even given a chance. We need to start a conversation about mucinous and other sub-types of ovarian cancer, so people understand the need.”**

**Sarah  
diagnosed at 25 with low-grade  
mucinous ovarian cancer**

Mucinous ovarian cancer is rare and can be difficult to diagnose. Mucinous tumours can be non-cancerous (benign) or cancerous (malignant). Clear cell ovarian cancer is rare and it can be linked to endometriosis. Endometrioid is the second most common type of epithelial ovarian cancer and can also be linked to endometriosis<sup>11</sup>.

## **Germ cell tumours**

These make up less than 2% of all ovarian cancers. Most ovarian germ cell tumours are benign but some are cancerous and may be life threatening. Ninety percent of

patients with germ cell tumours survive five years after diagnosis. Women in their 20s are more likely to develop this type of ovarian cancer<sup>12</sup>. The most common sub-types are teratomas, dysgerminomas, endodermal sinus tumours (also called Yolk-Sac tumours), and choriocarcinomas.<sup>13</sup>

- *Dysgerminoma* – germ cells that form sperm in males or eggs in females. The most common type of ovarian germ cell tumour but makes up only 1-2% of all malignant ovarian tumours.<sup>14</sup>
- *Yolk-Sac tumour* – cancers that begin in the germ cells that form sperm or eggs. They are the most common malignant germ cell tumour in children.

## **Ovarian stromal tumours**

About 1% of ovarian cancers are ovarian stromal cell tumours. More than half of stromal tumours are found in women older than 50, but about 5% of stromal tumours occur in young girls.<sup>15</sup>

## **Malignant (cancerous) Ovarian Sex Cord-Stromal tumours**

- *Granulosa cell tumours (CGT)* - The most common type of malignant stromal tumour is the granulosa cell tumour. This type of tumour is usually found in adults and GCT of the ovary cause higher than normal levels of estrogen in a woman’s body. The cause of CGT is unknown<sup>16</sup>. Adult GCT account for around 1% of all ovarian tumours<sup>17</sup>.
- *Sertoli-leydig cell tumours* - Sertoli-leydig cell tumours are cancers that begin in the female ovaries. The cancer cells produce and release a male sex hormone that can lead to the development of male characteristics such as a deep voice<sup>18</sup>. These are rare,

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accounting for less than 0.5% of all ovarian tumours. Although they can be present in women of all ages, they are most common in young women.

***“There are challenges and barriers to studying rare ovarian cancers. Because there are significantly fewer patients, it is more difficult to conduct clinical trials. This is compounded by less federal funding as well as less pharmaceutical funding to support the trials. The rare ovarian cancer community needs to be increasingly proactive in advocating through awareness, education, and making the case for cutting-edge preclinical research and innovative trials that will make a difference in the lives of women.”***

***David M. Gershenson MD  
University of Texas  
MD Anderson Cancer Center***

## CHALLENGES AND BARRIERS

While there is some research and a number of clinical trials being undertaken in rarer ovarian cancers<sup>19</sup>, there are significant challenges and barriers in advancing treatment. In the field of research, as there are fewer cases of rare ovarian cancer, it is harder to conduct clinical trials due to smaller patient populations, for example, more time needed to recruit for the trials<sup>20</sup>. For most countries, ovarian cancer is not a national health priority which often results in lower levels of government support for research, and lower industry funding<sup>21</sup>. Additionally, patients are faced with a lack of information about their disease and other unmet support needs. For example, when a woman is newly diagnosed, it is important that she receives expert advice from a gynaecological oncologist with expertise on their rare ovarian cancer.

## WHAT THE OVARIAN CANCER COMMUNITY CAN DO

As a global community we have a role to play to help raise awareness of rare ovarian cancers and highlight the gap in treatment options, research, and support for those impacted. This will help focus efforts across the advocacy, scientific and research community, policy makers and industry to work together to address this unmet need.

It is also important to showcase examples of good practice and where action has made a positive difference to patients, and continue to support our community by producing information that helps patients to better understand their disease and where they can get support.

Ovarian cancer research is generally underfunded in relation to the burden of

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the disease, rarer ovarian cancers being disproportionately impacted by this lack of funding. Those that fund cancer research should be encouraged to ensure rare ovarian cancers are included in their funding strategies.

## USEFUL RESOURCES AND FURTHER READING

The Coalition's [2022 Annual Virtual Global Partner Meeting](#) included a session on rarer ovarian cancers.

## SUPPORT

Many ovarian cancer patient organizations provide support services for all types of ovarian cancer. The World Ovarian Cancer Coalition has a list by country of our [advocacy partner organizations](#).

For rarer forms of ovarian cancer there are a number of organizations that raise awareness, provide support, or raise funds for research in this area. These include:

[Cure Our Ovarian Cancer](#) Based in New Zealand, Cure Our Ovarian Cancer is dedicated to leading change by raising awareness, supporting and advocating for women with ovarian cancer and funding life changing research. They have a particular interest in low-grade serous ovarian cancer and work globally to provide low-grade serous specific support, information, and fund world-leading low-grade serous cancer researchers to find a cure.

[STAAR](#) is a US-based non-profit dedicated to low grade serous ovarian cancer, co-founded by three women diagnosed with low-grade ovarian cancer who are passionate about extending women's lives via research for new treatment opportunities.

[The Inspire Ovarian Cancer Forum](#) has a low-grade serous ovarian cancer category.

[The Mucinous Ovarian Cancer Project](#) is a charity initiative dedicated to raising funds for mucinous ovarian cancer:

There are also a number of virtual support groups led by people with rarer forms of ovarian cancer. The following cover specific types of ovarian cancer:

- [Young women](#)
- [Low-grade serous](#)
- [Mucinous](#)
- [Borderline](#)
- [Germ Cell](#)

***“From a patient perspective, it is important that we see progress in this area and that rare cancers do get funding because that’s the only way that they get improvements in their outcomes”***

**Jane Ludemann  
Founder  
Cure Our Ovarian Cancer**

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