

## Rare and uncommon cancers

A rare cancer is one that occurs less than 6 times in 100,000 people and an uncommon cancer is found between 6 and 12 times in each 100,000 people. The problem with being rare is that there are not enough cancers to do the large trials comparing new treatments with old which are used to confirm the benefit of a new treatment in common cancers, so it can be difficult to determine how best to treat uncommon cancers. Unfortunately it is also the case that most of the available research funding is going to the more common cancers, so their outcomes improve whilst the rarer cancers have less treatment options. It is not surprising that rare cancers are responsible for almost one third of the deaths from cancer.

We may soon, however, be able to learn from the common cancers how best to treat rare cancers. Up until now, we have determined the type of cancer by which of the body's organs it most resembles under a microscope. Now we are able to find what genes in the genetic material in the centre of the cancer cells have changed in order to trigger the cancer. These changes and the proteins made from the altered genes can be targeted by drug therapy or immune therapy. It has been found that cancers which look different can share the same gene alterations and therefore may respond to similar treatment. Likewise, cancers which look the same can respond differently because they have different patterns of altered genes.

Ovarian cancer, for example is uncommon. It is difficult to detect because there are no screening tests (unlike Pap smears for cancer of the cervix) and so the ovarian cancer can be quite advanced when it is diagnosed. The early symptoms are also vague. Fatigue, stomach and back pain, bloating and menstrual irregularities can mimic many other conditions. More than that, ovarian cancer already has several different looking subtypes which respond differently to treatment and not surprisingly different patterns of altered genes which also respond differently to each other.

Around one in six ovarian cancer have altered BRCA 1 and 2 genes the genes commonly altered in inherited breast cancer. People with these altered genes may be able to prevent developing ovarian cancer by removal of the ovaries and tubes before cancer develops, or at least be closely watched for early cancer.

In addition it has been discovered in breast cancer that a new class of drugs, the PARP inhibitors, work in a particular type of breast cancer. These drugs are now being trialled in a specific type of ovarian cancer which shares the same genetic alterations. In future, instead of doing large trials comparing one treatment to the other in rare cancers or rare subtypes of common cancer we may firstly need to show that a drug which targets the altered gene or its protein is effective in a small study, start using it widely, and then by combining cases treated across the world, refine the treatment as we discover the characteristics of the patients who respond the best.

By finding the genetic similarities between rare and more common cancers we may be able to make gains in the treatment of rare cancers.

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## **Publication**

[Reassessing rare cancers.](#)

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