Cancer: it's not all about the Big Four

There are 200 cancer types that few have heard of, yet they make up half of our cases each year, says Simon Crompton

Myelodizzy what? No one in America, apart from those who had treated, studied or experienced it, knew about a rare blood cancer called myelodysplastic syndrome (MDS) — until last month. That was when Robin Roberts, popular host of ABC's *Good Morning America*, tearfully announced to the nation that she was undergoing treatment for the condition, and would be out of action for a while. Now everyone knows about MDS: websites offering information are crashing and potential bone marrow donors are rushing forward.

In the UK it's a little different — and not just because we emote less on morning television. Many doctors haven't heard about MDS, there are no official figures on how many people have it (it's probably around 7,000), the MDS charity has two part-time employees and an annual income of just £60,000 (compared with Breakthrough Breast Cancer's £18 million), and finding corporate sponsors and money for new treatments is difficult. This isn't a one-off. There are more than 200 types of cancer, many with long names of which few people have heard, many represented by small charities shouting "What about me?". However, the big four cancers — breast, bowel, prostate and lung — dominate national interest, media profile and charitable giving.

These "rarer" cancers make up half of all cancer cases in the UK, affecting hundreds of thousands of people. But survival rates are generally worse than the most common cancers. Just 47 per cent of people with rarer cancers survive for five years compared with 77 per cent for prostate cancer and 85 per cent for breast cancer.

Why? It's partly a reflection of the fact that rare cancers can often be particularly nasty ones that tend to be diagnosed late, and because they tend to get little publicity and (individually at least) don't affect many people. That means funding for research and new treatments — even for cancers of the pancreas, throat, stomach, bladder and ovary, which aren't the most common, but aren't rare — is harder to obtain.

It's bad enough receiving any diagnosis of cancer, but harder when you discover that treatments, expertise and support are hard to find. At least the nurse giving Nicola Wardle her diagnosis over the phone was honest.

"She said I had a cancer called a gastrointestinal stromal tumour (GIST), and that in 30 years of experience she had never heard of it," says Wardle, 43, from Leeds. "It's been the same with doctors I've spoken to. You just feel so isolated and freakish, and as ask yourself 'why me?'"

There are only 200 to 900 new cases of GIST in the UK every year (compared with 41,000 cases of lung cancer), and it has only been possible to diagnose the condition unambiguously since 2000. After diagnosis, the doctor provided Nicola Wardle, a former teacher, with no information apart from the news that her case was unusual and she needed more surgery. She recalls her husband having done an internet search, face white as a sheet: he'd found some facts. Only half of those who had a GIST removed survive five years. That was five years ago.

It was only when she started to attend a GIST Support UK group that Wardle, who has sons aged 7 and 11, began to feel human again. She feels no resentment against the high profile of other cancers, and has raised thousands of pounds for Cancer Research UK, knowing that it funds research into rare cancers. No one denies that the bigger cancers need the money too, but what frustrates Wardle is that there are so few clinical trials investigating new treatments for GIST. "There's so much red tape that gets in the way of smaller research projects," she says.

Judy Dewinter, 47, has had similar experiences. A city high-flyer with her own brokerage business, she was diagnosed with myeloma 14 years ago. She tried to show no weakness in the City's cut-throat environment. But in 2003 she had to "come out" when she needed a bone marrow transplant. She gave up her job, and has spent her time since then bringing up her family and trying to raise funds for a disease that makes up 1 per cent of all cancers.

"Myeloma's not all that uncommon, but have you ever heard of it?" she says. It affects bone marrow cells, crippling their ability to produce infection-fighting antibodies. Around 37 per cent of people with the condition live for five years. As a fundraiser, Dewinter is trying to convince people of the relevance of rarer cancers to all of us. If estimates suggest that four in ten of us will get cancer, then 20 per cent of us are destined to have one of the less common varieties. Tiny charities such as Myeloma UK, she says, are trying to plug the gaps that the Government, the NHS and drug manufacturers are not filling.

"It's very frustrating for patients in the UK because you can see that there are trials and new drugs available for your type of cancer in other countries, but not here," Dewinter says.

Myeloma UK is among the organisations that have tried to persuade the body that rations NHS treatments, the National Institute for Health and Clinical Excellence (Nice), that it cannot apply the same rules to common and uncommon cancers.

It started negotiating with Nice when people with myeloma in the UK became aware of the life-transforming drugs available in America and other European countries, but not here. A drug called Velcade is one example. Trials showed that the drug could extend the lives of people with myeloma by as many as seven years, but the drug company priced it high — £9,000 to £18,000 per course of treatment — because it would only be used by a few hundred every year.

According to Professor Gordon Cook, a consultant haematologist at Leeds Teaching Hospitals and a director of Myeloma UK, the price was too high to pass Nice's strict formula to determine if a drug's cost was justified by its benefits. Then Myeloma UK and other bodies started negotiations with pharmaceutical companies, so that they effectively reduced the cost to the NHS. Nice relented, and the model has been repeated with other drugs for rare cancers.

There are now signs that government, the NHS and research charities are waking up to the problems posed by rarer cancers. Six years ago, an organisation representing 50 organisations for less common cancers was set up at the request of the Government's cancer czar, Professor Mike Richards, to have input into the national cancer reform strategy. It was named Cancer52, after the percentage (at that time — it's now 53 per cent) of deaths to rarer cancers. Since then, by providing a strategic alliance of smaller charities, it has given them considerably more clout in influencing the NHS and research agenda.

Cancer Research UK is keen to emphasise that many of its initiatives make rarer cancers a priority, and the perception held by some with rarer cancers that it concentrates research funds on the more common cancers is wrong. Late last year it announced that it would join with research groups in the UK, Europe and the US to design and fund large trials into new treatments for rare cancers (the International Rare Cancers Initiative).

Kate Law, Cancer Research UK's director of clinical trials, says that Cancer Research UK will be providing infrastructure for small research teams, helping them to cut through red tape, as well as providing funding.

She senses a change in mindset in pharmaceutical companies too: although they may not want to run trials on their drugs for rare cancers themselves, several have expressed an interest in providing drugs, logistics and information to support Cancer Research UK-funded academics who want to research them.

Andrew Wilson, the chief executive of the Rarer Cancers Foundation, believes that the real challenge is to deal with the lack of awareness in health professions. Late diagnosis is a major problem.

"GPs need to start thinking out of the box, so if symptoms can't be explained by common illnesses they refer to specialists earlier. Cancer nurse specialists need more education about rarer cancers," he says.

Rare cancer organisations

Cancer52 cancer52.org.uk
GIST Support UK gistsupportuk.com
Myeloma UK myeloma.org.uk
MDS UK Patient Support Group mdspatientsupport.org.uk
Rarer Cancers Foundation rarercancers.org.uk

You've got what? Rare cancers explained

Adamantinoma One of the rarest bone cancers
Cholangiocarcinoma Bile duct cancer
Dermatofibrosarcoma protuberans Very rare, lump-forming skin cancer
Esthesioneuroblastoma Rare tumour of the nasal cavity
Gigantiform cementoma Rare tooth tumour
Hairy cell leukaemia Type of cancer of white blood cells (which have hair-like outgrowths)
Juxtaglomerular cell tumor Rare kidney cancer causing high blood pressure
NUT midline carcinoma Aggressive head and neck cancer caused by changes in the NUT gene
Oligodendroglioma Type of brain tumour arising from cells called oligodendrocytes
Waldenström's macroglobulinemia Cancer of the immune system, causing overproduction of an antibody

ADAM GERRARD, AND BEN GURR

Judy Dewinter, who suffers from myeloma, and, left, Nicola Wardle, who has a rare gastrointestinal stromal tumour