

Sarcoma UK

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Email: info@sarcoma-uk.org

Welcome

If you've picked up this newsletter it is probably because you, a member of your family, or a close friend has been diagnosed with sarcoma. If the first diagnosis was sometime ago one of the first things you may have discovered, just as I did, was that finding out about sarcoma isn't easy.

I was first diagnosed in 1999 with a soft tissue sarcoma. I had treatment over 18 months before its progress was halted and I have now been free of disease for over two years. I have had a lot of help during that time and Sarcoma UK is an attempt to pass on some of the things I have learned in a practical way. The aim is to offer information and ideas, give news about how sarcoma treatment is developing and to give you an opportunity to make your voice heard too – I welcome any letters or emails.

I should like to thank the wide range of people who have been encouraging me to start this newsletter, the specialist oncologists and surgeons, nurses and other patients, and also those who have commented and contributed during its development.

Please write – I look forward to hearing from you.

Roger Wilson

Developments in Sarcoma Treatment

The latest developments in the sarcoma treatment were discussed at a one day conference in London hosted by the Royal Marsden Hospital in February. Many of the leading surgeons and oncologists treating sarcoma patients were there and joined in discussions on various themes ranging from diagnostic practice, to the latest developments in chemotherapy and to the organisation of specialist sarcoma centres.

The average size of a sarcoma tumour is 10 centimetres. That's about the size of a tin of beans, explained Mr Rob Grimer, surgeon oncologist at Birmingham's Royal Orthopaedic Hospital. This compares with an average of 2.5cm for breast cancer tumours. The breast cancer screening programme is a major contributor to early diagnosis and means that there is a positive outcome for most people facing the disease. Getting an early diagnosis is just as crucial for sarcoma as it is for breast cancer but similar screening for sarcoma is impossible.

The relationship between advanced disease at diagnosis and the size of

the primary tumour is clear and it is known that the long term survival of those with a smaller tumour is generally better. The sarcoma guidelines followed by GPs for referring a patient to a hospital consultant uncover fewer than half of all diagnoses of soft tissue sarcoma. The remainder come from referral for some other suspected complaint and patients often have surgery before the true nature of their disease is discovered.

Professor Ian Judson (medical oncologist at the Royal Marsden Hospital) reviewed recent developments in chemotherapy. He commented that ten years ago there wouldn't have been anything much for him to talk about. Patients needing chemotherapy then had few options. Combinations of new drugs were showing promise in clinical trials and although overall progress was slow patients were living longer than they otherwise would have done. New drugs are now arriving. Among the most promising in recent trials is Yondelis (trabectedin, ET743), which is currently being evaluated by the European medicines regulator, EMEA, for

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advanced soft tissue sarcoma. The star performer amongst the new drugs is Glivec (imatinib). Although this is specific to gastro intestinal stromal tumour (GIST) it shows the way for new drugs designed to address specific tumour characteristic.

The work of the Royal Marsden's Rehabilitation Ward (the Markus Ward) was described. Patients undergo a full assessment before their surgery so that their recuperation can be programmed with their lifestyle, wishes and needs in mind. The importance of understanding that patients can feel isolated by a rare cancer, are well understood and a wide range of multi-disciplinary support skills is available.

Dr Anna Cassoni (clinical oncologist at University College Hospital London) described the role of radiotherapy in supporting surgery as the primary curative treatment for sarcoma. Not every patient will benefit from radiotherapy but where it is used the liaison between surgeon and radiotherapist needs to improve, she said. There is a need for clinical trials to address some of the issues of dosage and patterns of treatment because a good proportion of patients would benefit.

Mr Matthew Clark (surgeon at Queen Elizabeth Hospital Birmingham) reviewed the complexity of the major surgery which large sarcoma tumours involve. Patients, surgeons and their support teams face severe challenges when tumours get very large and he made the point that however advanced the surgical techniques, the overall care and support needed by the patient must be every bit as advanced.

NICE Appraisal for Glivec

The government has asked the National Institute for Clinical Excellence to conduct a technical appraisal of Glivec for treating GIST. This is a key step in the process of deciding whether Glivec should be made available through the NHS to all the patients that doctors think should take it.

One of the characteristics of GIST is that it is resistant to treatment. International trials giving Glivec to patients with advanced disease started in 2000 with startling results and it has been one of the fastest ever drugs through the approvals process.

Sarcoma UK has been invited to make a submission to NICE for consideration by the Appraisal panel. We would like to hear from any GIST patient or carer, whether on Glivec or not, so that we can prepare a submission which truly reflects what patients think and feel about this new drug.

Please contact us by email (info@sarcoma-uk.org) or mail to PO Box 5, Church Stretton SY6 6ZZ.

GETTING INFORMATION

The first stop for information about your case and your treatment should be your doctors. The hospital specialist will have the most information and will be able to tell you most about the treatment appropriate for you. Your GP will have some information but remember that on average a GP will only ever have one sarcoma patient in his whole career! Sarcoma is that rare!

General sarcoma information is available from Cancer Bacup. They have good booklets on Soft Tissue Sarcoma and on Understanding Primary Bone Cancer. Your hospital information centre may have them in stock but if not call Cancer Bacup on the publications phone number below and they will post them to you straight away. Some hospitals also have local information in their information centre.

If you have access to the Internet there is a wide range of information available, from around the world. Even if you cannot access the Internet yourself there may be someone who can do it for you and print out the pages they find. It could be quite a lot of paper !

Information can help you decide what questions you want to ask. Doctors or nurses don't mind being asked, they would rather hear all the questions you want answered and they will spend time answering them. Their focus is on you, the patient.

If you have been diagnosed with advanced sarcoma information is just as important as it is for the newly diagnosed, though it may be of a different kind. Treatments such as chemotherapy have a lot of mystique about them and raise fears of their own. Rather than listening to old stories about side effects it is much more valuable to hear about how treatments have changed and how modern medicine, and some complementary therapies too, can help you through a demanding treatment.

It is also important to understand that if you do not want to learn too much about your disease you don't have to. The choice is yours.

CONTACTS

CANCER HELP UK

Cancer Help is Cancer Research UK's patient website. Excellent sarcoma material including "Questions for Your Doctors". Click the link to 'Specific Cancers' and also use the search function as there are further references within the site.

<http://www.cancerhelp.org.uk/>

CANCERBACUP

Cancer Bacup's free helpline is staffed by experienced nurses who can answer many questions. The web link leads to booklets on sarcoma which are available free by phoning Bacup. There are many other booklets covering all aspects of cancer.

<http://www.cancerbacup.org.uk/catalogue/sarcomas.htm>

0808 800 1234 - freephone helpline

020 7696 9003 - publications

MACMILLAN

Macmillan works to improve the quality of life for people living with cancer. It offers information and practical advice over the phone and its web site is extensive. Macmillan has information on local support groups.

<http://www.macmillan.org.uk/>

0800 808 2020 - freephone helpline

UK CHILDREN'S CANCER STUDY GROUP

The UKCCSG is a main contact point for professionals treating children's cancer. Support materials and downloadable newsletter are on the website.

<http://www.ukccsg.org/>

SARCOMA UK

We are a new publication so our web presence has yet to grow. The main page offers links to a wide range of further Internet resources which is growing regularly.

<http://www.sarcoma-uk.org>

PHARMAMAR

Pharmaceutical company with good information on sarcoma.

<http://www.pharmamar.com>

Be cautious when using the web to look for information on any cancer. Just because a website exists it does not mean that those responsible have any expertise. Information can only be general and can not be specific to a particular patient.

HOW MANY CASES?

The incidence of gastrointestinal stromal tumours (GISTs), a rare sarcoma, may be more than three times as high as previously believed, according to a study from Sweden. An accurate estimate of GIST incidence has been elusive because of the difficulty of accurate diagnosis. The availability of an effective therapy makes it vital that diagnosis is accurate. The higher incidence of GIST is calculated at 16 per million people annually.

The Swedish researchers analysed medical records and tissue blocks of patients seen between 1983 and 2000 and found 400 cases, more than three times as many as had been originally diagnosed. Many of the patients still living, and whose diagnosis was changed, are now candidates for treatment with Glivec, the drug which has revolutionised GIST treatment.

Annual incidence of 16 per million suggests about 800 new GIST cases per year in the UK. Previous estimates suggested an annual occurrence of less than 200 but it is known that such figures have been never been accurate.

It sounds astonishing but it is true. No-one is certain how many sarcoma cases are diagnosed each year in the UK. These days people are hungry for statistics but the fact is, the National Health Service does not gather data about sarcoma in a way allows an accurate picture to be built up.

Cancer Research UK published figures for 1998 which show 1312 soft tissue sarcomas diagnosed and 461 bone sarcomas. That makes sarcoma very rare indeed, under 1% of all cancer diagnoses in the UK.

There is evidence though that these figures may under-estimate the total quite substantially. Why, seems a fair question to ask.

The data on cancers are gathered by cancer 'site' - for example, breast, prostate or colon are cancer sites. Sarcoma is cancer of 'connective tissue' and because such tissues are everywhere in the body, sarcoma can appear almost anywhere. Although the majority of cases are well defined, being on legs, arms or in bones there are a lot of cases of internal sarcoma, or sarcomas attaching themselves to specific organs in the body, all of which means that cancer 'site' specialists can end up finding them and treating them along with their regular caseload.

The difficulty of diagnosing sarcoma is another problem. Because these cancers are so rare many doctors see very few cases and the number of pathologists who see sufficient cases to be sure of diagnosing tumours accurately first time is small.

The development of new reporting systems in the NHS and a better database for gathering them will hopefully improve our understanding, and our doctors' understanding, of sarcoma. Even if the numbers rise because of better systems it will not mean there are more cases, just that the cases which went unreported are now being spotted (see box for research on statistics.)

If that means that patients get better treatment the work will have been worth it..

GETTING THE DIAGNOSIS

Getting to a diagnosis is often the first hurdle facing a soft tissue sarcoma patient. The rarity of these tumours and the inexperience of most doctors in identifying them mean that mistakes are made and quite often patients have surgery which is inadequate.

The situation is a little better where there is a regional centre specialising in sarcoma treatment. In the major cities sarcoma centres support patients from a wide surrounding area and in London the Royal Marsden Hospital treats patients nationally.

However in much of the UK it is a lottery. Many parts of the country have no recommended referral route for suspected or diagnosed patients and as there are no national guidelines on treatment there are no consistent standards of care.

Sarcoma UK is conducting research which we hope will contribute to the understanding of this problem and speed up improvements.

WHAT IT MEANS

The Multi-Disciplinary Team

Sarcoma can be a difficult cancer to diagnose and because it is so rare it is very unlikely that sarcoma will be considered as a diagnosis in early consultations with doctors.

On average GPs will only encounter one soft tissue sarcoma patient during their whole career. Many surgeons have never seen a sarcoma patient and most cancer specialists have little experience of treating them. Although diagnosis and initial surgery may be undertaken locally, follow-up treatment should be decided by those experienced in dealing with sarcomas and is usually managed through a specialist centre, or in consultation with a specialist centre. When disease is advanced at diagnosis, or metastasises later, the specialist treatment centre will have access to the latest treatments and may also be able to access clinical trials, which are used to test new drugs and treatments.

The specialist centres operate a team approach to treatment. Surgeons, oncologists, radiographers, pathologists and other specialities form a 'multi-disciplinary team' and will discuss the most appropriate way of treating each patient at the key stages in their illness. There will be nurses and other support staff too with specific experience of treating sarcoma patients. The multi-disciplinary team at the specialist sarcoma centre offers patients the best survival chance.

You may only ever meet one medical member of the team at a clinic, and sometimes that may be a junior member still undergoing training, but they will be reporting to an experienced consultant and treatment decisions or recommendations will be made only after all the options have been considered.

Sarcoma is survivable. A large proportion of patients have good quality surgery, need no further treatment, and never have another problem. Even so the doctors will monitor patients regularly because this is a cancer which does recur, without warning, and sometimes after many years in remission. No-one knows why and trying to find out is the subject of research here and around the world.

ONCOLOGIST

A doctor who specialises in the treatment of cancer. You will find medical oncologists, clinical oncologists and surgeon oncologists.

Medical oncologists specialise in the use of drugs (chemotherapy) while clinical oncologists are specialists in using radiotherapy.

CLINICAL TRIALS

The UK's national cancer research programme calls for an expansion of the numbers of patients joining clinical trials. Many hospitals will be running trials for the very first time but for rarer cancers, like sarcomas, most trials are likely to be found in the main treatment centres under the guidance of surgeons and oncologists who specialise in treating sarcoma.

The purpose of clinical trials is to test and evaluate new treatments, drugs and procedures to assess whether they should become the standard treatment for the situation in which they are being trialled.

Phase I trials, usually of new drugs, enlist very few patients and are designed to establish what the tolerable and safe dosage is.

Phase II trials recruit a larger number of patients (rarely more than 40 to 50). These trials test the effect a drug has on the target disease in various dosages, patterns of treatment etc.

Phase III trials recruit larger numbers (often several hundreds) and are randomised comparisons between existing and new treatments. All patients are randomly selected, some for the standard treatment and some for the experimental treatment.

During trials individual patients are carefully monitored and if one treatment is clearly better than the other, or if the experimental treatment shows no benefits, the trial can be stopped. Those joining a trial can always choose to leave it and receive the standard treatment.



JANINE'S SEXY LEG



BBC Radio Newcastle presenter Eddy Secker with Janine Broadbent (right) following her radio appeal.

“I know money won't bring my leg back, but in the long term it might just stop someone else losing theirs. I strongly feel that, especially with such a low-profile disease such as this, a little money could make a huge difference.”

I was just 24 (I'm now 27) when I was diagnosed with an osteosarcoma. Albeit I was rather 'old' for this disease, age makes little difference to the impact it has on your life and how alone you feel going through treatment and then learning to live with its consequences. Just three months before, I had completed my degree in Management and I'd just been accepted for a job with British Airways as a stewardess.

After many months of chemotherapy I had to lose my leg above the knee and now I walk using an artificial leg.

I would love to be able to wear skirts and sandals comfortably again but unfortunately life-like limbs are very expensive. With the encouragement from two nurses, now very special friends, Claire and Kirsty, I set up the 'Help Get Janine A Sexy Leg Fund' in the hope of raising funds for a new sexy leg! They reckoned I could go on proper nights out with them if I could wear a skirt and sandals, instead of the trousers and boots I've had to make do with since November 2000.

Fundraising has been extremely successful. We've held many events, and the support I have received both locally and nationally has been overwhelming. The name of the fund reflects the way I feel about my situation. I'm very positive despite what has happened to me and I want to make people around me smile and see the bright side. The 'Help Get Janine A Sexy Leg Fund' has certainly grabbed everyone's attention. I have been asked to give motivational speeches, featured in both local and national newspapers and also interviewed on BBC Radio Newcastle!

But raising money for my leg is only the start of what I consider my real mission. That is to increase awareness of and funding by setting up a sarcoma charity. I know money won't bring my leg back, but in the long term it might just stop someone else losing theirs. I strongly feel that, especially with such a low-profile disease such as this, a little money could make a huge difference. If we look at just how far sarcoma treatment has advanced in the last 30 years or so with relatively little funding in comparison with the other commoner types of cancer.

I've also become involved with the Sarcoma Group and the Consumer Liaison Group within the National Cancer Research Institute as a patient representative so I now have access direct into the heart of current UK sarcoma research.

We've also just established a sarcoma support group which meets once a month in Newcastle. We call ourselves 'Sarcoma Survivors' and provide peer support, communication, education, and information with guest speakers and helpful, wide-ranging discussion topics.

I'm really excited about the future and I'm very passionate about what I'm doing. My degree gave me great back-up skills and I'm sure they will come in very useful. The Help Get Janine a Sexy Leg Fund is hopefully just the start. I feel I could devote the rest of my life to helping lift public awareness of sarcoma and the victims of the disease.

If you would like to help Janine get her new leg (or make a donation to sarcoma research) you can contact the fund via email at helpgetjanineasexylegfund@btopenworld.com

SARCOMA RESEARCH

Research into sarcoma in the UK is managed by the National Cancer Research Institute (NCRI). The NCRI is a partnership between the government and the major cancer research charities with the ambition of making UK cancer research a world leader once again.

There are two main areas of research. The first is into the genetics of cancer and here work is not sarcoma specific. The second area principally concerns new treatments and although both surgical and radiotherapy techniques are developing the most significant activity concerns chemotherapy. Pharmaceutical companies are important partners – they invest billions each year on research into new drugs, although new drugs to treat sarcoma tend to result from research into other cancers.

It can take many years for a new drug to reach the stage of entering clinical trials and then to gain full approval for general use. Even then trials continue as new patterns of treatment and combinations of different drugs are tested. Developments in recent years mean that

the range of drugs with proven value against sarcoma is slowly increasing, although many will not be in general use for some years.

Clinical research – trials run with patients in hospital – is co-ordinated through the NCRI Clinical Study Group for sarcoma which is responsible for oversight of clinical trials in the NHS. Because sarcoma is so rare research relies on collaboration across Europe as well. The European Organisation for the Research and Treatment of Cancer (EORTC) has a Soft Tissue and Bone Sarcoma Group which is recognised worldwide as the leading international research group in sarcoma. EORTC sponsors large scale trials which cannot take place in the UK alone because the number of eligible patients is so small.

We look briefly at four of the newer drugs – two of which are only just embarking on their journey towards approval. There is no guarantee they will make it – that's the nature of this research.

GEMZAR

Gemzar, also known as Gemcitabine, went into clinical trials in the USA in the early '90s for the treatment of pancreatic cancer. It was approved in 1996 and then in 1998 also received approval for lung cancer patients. It is given intravenously.

Gemzar is being studied in a variety of tumour types including sarcoma. It is showing promise as a second line of treatment when being given in combination with other drugs. In one trial given with docetaxel it attained a 53% response rate for patients with uterine leiomyosarcoma.

YONDELIS®

Yondelis® (trabectedin, ET-743) is derived from a Caribbean sea squirt – a primitive sea creature. Developed by Spanish company Pharmamar it has shown remarkable activity in Phase II clinical trials against soft tissue sarcoma. Patients with advanced disease have had extended progression free and overall survival.

Yondelis® has a distinct multidirectional mode of action that inhibits cell proliferation, leading to cancer cells dying. It has been granted 'orphan medicinal product' status for the treatment of soft tissue sarcoma by the European Commission which should lead to its rapid approval for clinical use. Yondelis® is currently under review by EMEA for advanced soft tissue sarcoma.

BROSTALLICIN

Brostallicin (from Pharmacia Corp) binds to base pairs in DNA preventing cell reproduction. It is already in Phase II trials in Europe and the USA for a wide range of cancers including advanced and metastatic sarcoma.

SUGEN SU011248

This so far unnamed drug is designed to damage tumours by attacking the blood vessels that 'feed' them. It had good response in its first patient study, in France, with patients suffering from a range of advanced cancers, including angiosarcoma. Phase II trials are starting in 2003.

Sarcoma UK is supported by an unrestricted educational grant from Pharmamar.

If you require additional copies of Sarcoma UK for distribution through hospital, support group, charity etc please advise us by email of the number of copies required and the mailing address.

Email to: info@sarcoma-uk.org

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TELL US YOUR STORIES

The UK's small community of sarcoma patients has never had the opportunity to exchange views and experiences before.

Tell us your stories, talk about the bad moments and the good. Share your laughter as well.

I lay on the bed as the nurse put on her plastic apron and gloves before changing the bag of chemo attached to my drip. I asked why she had to wear protective clothing. She looked at me seriously, but with a twinkle in her eye.

"I mustn't let this stuff get on my skin you know, it could cause cancer".

We both burst out laughing. She was, after all, pumping it into me.

Editor's own true story

If you want to receive future copies of Sarcoma UK by post please complete the details below and post the form to: Sarcoma UK, PO Box 5, Church Stretton, Shropshire, SY6 6ZZ. Alternatively email us at info@sarcoma-uk.org.

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Please tell us a bit about yourself by ticking appropriate boxes below. **Are you:**

- | | | | |
|-------------------|--------------------------|-----------------------|--------------------------|
| sarcoma patient | <input type="checkbox"/> | other nurse | <input type="checkbox"/> |
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| friend | <input type="checkbox"/> | other NHS | <input type="checkbox"/> |
| GP | <input type="checkbox"/> | local support group | <input type="checkbox"/> |
| cancer consultant | <input type="checkbox"/> | national organisation | <input type="checkbox"/> |
| other doctor | <input type="checkbox"/> | commercial company | <input type="checkbox"/> |
| cancer nurse | <input type="checkbox"/> | other | <input type="checkbox"/> |

We are aiming to set up an **email support group**. If you have an email address and would like to be contacted about joining such a group please tick.

Where did you get your copy of Sarcoma UK?

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| other | <input type="checkbox"/> |