

Sarcoma UK

Sixth Edition
Spring 2006

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Inside:

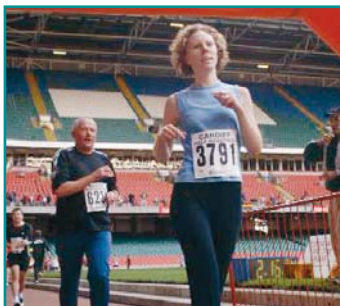
Scientists working on sarcoma research

See page 2



The wrong time ... a challenging memory

See page 6



A big thank you to our supporters

See page 7

CAMPAIGNING FOR PROPER TREATMENT

Sarcoma UK is campaigning for changes to the NICE guidelines for the treatment of inoperable or metastatic gastrointestinal stromal tumour (GIST). GIST is one of the more common sarcomas and affects the stomach and intestines. In 2000/01 the new drug imatinib (Glivec®) transformed the treatment of this sarcoma and patients with a very poor prognosis were offered a new lease of life.

However resistance to the treatment builds up over time and the NICE Guidelines, published in 2004, have been overtaken by significant new research which means that the treatment they prescribe is now inappropriate.

The situation is very different from the public debate about Herceptin for breast cancer. The patients affected have active disease, there is appropriate treatment, but NICE guidance allows Primary Care Trusts to refuse to fund it. The NHS allows patients to appeal against these decisions and Sarcoma UK has been supporting patients as they seek funding. A number of appeals to PCTs have been won.

These appeals have been helped by the work of a consensus group of specialist surgeons and oncologists who treat GIST. They came together in the autumn of 2005 to develop an evidence-based set of guidance on treatment of the disease. This authoritative document is now followed in most treatment centres and is increasingly being recognised by PCTs to whom it has been given.

Working with the specialist doctors and other patient groups, including GIST Support UK and Cancer BACUP, we are preparing a petition to the Department of Health to have the NICE Guidelines set aside so that clinical decisions can be made on the basis of patient benefit rather than be driven by funding pressures.

Mike Scott won funding from Morecambe Bay PCT for an escalated dose of imatinib to be made available for him should he require it, following an appeal against the PCT's first decision supported by his doctors at the Royal Marsden, his MP, his GP and Sarcoma UK.

Mike, who lives in Barrow-in-Furness in Cumbria, has faced extensive surgery for his GIST, a year of imatinib at the standard dose, and is currently taking sunitinib (Sutent®) on an extended clinical trial. Should he relapse with this treatment he now has access to imatinib at an escalated 800mg dose.

His story made front page news in Cumbria and Mike hopes that his success will give inspiration to others to challenge the system and win appropriate treatment.



Also Inside:

Patient Story
Raising Awareness
Latest information on clinical trials
Getting more information – see back cover

Sarcoma UK

Sarcoma UK is supported by unrestricted educational grants from Novartis Oncology and Pharmamar SA.

This newsletter is published free of charge to sarcoma patients, carers, friends and those who treat this group of rare cancers.

If you require additional copies of Sarcoma UK or would like to distribute it through hospital, support group, charity etc please let us know by email of the number of copies required and the mailing address.

Email to: info@sarcoma-uk.org or to sarcoma@dflair.demon.co.uk

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Editor Roger Wilson

Design Chris White

If you would like to join our email support group please contact us at:

info@sarcoma-uk.org or go to www.mailtalk.ac.uk/lists/sarcoma.html

We are keen to hear and publish patient stories. Stories we have printed already have proved an inspiration to other patients. Tell us about your sarcoma experiences.

Address:

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EUROPEAN SARCOMA RESEARCH COLLABORATION

Sarcoma's rarity is a great hurdle in the fight to understand this disease and to find and access better treatment options. Laboratory and clinical research both need large numbers of patients for the evidence gained to have significant impact. This means that there need to be large numbers of tumour samples for laboratory use, or patients prepared to enter clinical trials - something that is almost impossible for individual research institutions or hospitals to achieve.

To overcome this, a group of internationally eminent sarcoma researchers from across Europe who work in academic research institutions and hospitals (including the Institute of Cancer Research in the UK) have come together to form a sarcoma research consortium called CONTICANET (CONnective Tissue CANcer NETwork). CONTICANET is also being supported by pharmaceutical and biotechnology companies.

The aims of CONTICANET are ambitious and wide-ranging but its principal task is to more effectively study the underlying biology of sarcoma and other connective tissue tumours. The aim is to develop more effective therapies by bringing together this critical mass of researchers, resources and knowledge and by integrating research projects across Europe through the consortium.

The importance of patients and their carers cannot be underestimated, and their participation is seen as vital to the success of this consortium. Patients can donate tissue and consider entering clinical trials if available, but can also play an important part through helping to guide the direction of the consortium, and by lobbying and disseminating information. Sarcoma patient advocacy groups are already involved, including Sarcoma UK, and have helped to nominate members of the Scientific Advisory Board. Plans are in hand for a patient advisory committee within the next 12 months.

CONTICANET also sees one of its roles as providing a platform for advocacy groups from around Europe to come together, as well as providing support to disseminate information from patient groups.

CONTICANET has won a 5 year European Commission grant that will allow the integration of the research groups involved. On the 1st/2nd February this year members from all of the participating institutions met in Paris for the inaugural meeting and the work began enthusiastically and in earnest from day one. Working groups have been set up to deal with the various integration issues so that research can move forward quickly. There is much to be done and everybody is working hard to make this initiative a success for all of us in the sarcoma community.

With thanks to Dr Michelle Scurr for this report

GUIDELINES PUBLISHED

The National Institute for Health and Clinical Excellence published the service guidance for sarcoma treatment in the NHS in March. The full documentation from the work, which took two years, can be downloaded from the NICE website at <http://www.nice.org.uk/page.aspx?o=297430>

The key recommendations are:

- Prompt referral for expert diagnosis is crucial
- People should be treated by a specialist multidisciplinary team
- Treatment should be carried out by specialists
- Appropriate support and rehabilitation services should be available to people who are disabled by treatment for sarcoma
- All sarcoma teams should collect data on treatment and care, and take part in training programmes and audits.

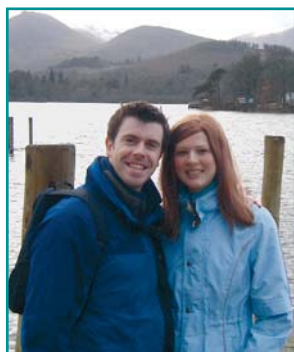
PATIENT STORY

Sally Young is a local radio journalist with the BBC and found herself challenged in ways that few of her colleagues can imagine. She shares her story.

Twelve months ago I was a 26-year-old journalist, passionate about salsa dancing, and the man I was about to marry. The only worry I had was a persistent knee pain that was getting steadily worse, despite numerous visits to the doctor. He put it down to a strain from my dancing, but when my knee became swollen and red and I could barely walk I insisted on an x-ray. From the moment the x-ray film was produced, things started moving very quickly and I was sent down to the Royal Orthopaedic Hospital in Birmingham for a biopsy. Three weeks later, but six months after I'd first gone to my GP, I was diagnosed with osteosarcoma: bone cancer.

My first fears on hearing the diagnosis weren't about my mortality or hair loss from chemotherapy or the impending operation to remove the diseased bone; they were about my fertility. I'd always known I wanted to be a mum and I also knew that chemotherapy could cause infertility. I didn't want to start treatment until I'd saved some eggs. But then came the second blow: if I underwent fertility treatment now, my chances of survival would be drastically reduced. I made the difficult decision to start chemotherapy straight away. I don't think anyone around me could understand my feelings at this time. My fiancé Pete, my parents and my friends just wanted me to get better. And of course rationally, they were absolutely right: what was the point of being able to have children if I didn't survive? But somehow it didn't feel like that to me then, and today I still face the pain of my probable infertility every day.

But I had to put the worries over my fertility to one side while I underwent chemotherapy. I had expected it to be tough, but I was almost overwhelmed by the treatment at the start. After my first five-day hospital stay I returned home in a wheelchair, unable to even to watch television or read a book, let alone make a meal or climb the stairs. I felt as if I was neither awake nor asleep, but in a kind of purgatory. Meanwhile, Pete was having to cancel our wedding plans. I think this was my

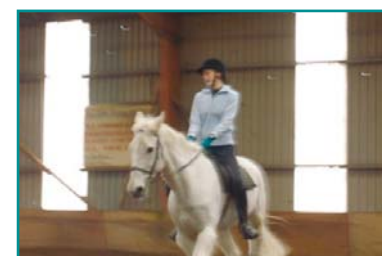


lowest point: I felt as if I would rather die than continue with this treatment for nine long months. But I did get through the days, by relying on my family and friends to care for me.

Two months into the chemotherapy treatment, I had a scan on my knee. To my horror, the tumour had actually got bigger and I was told my best chance of survival would be to have an amputation. How can you prepare yourself for an operation like that? Once again there wasn't time to think about things too much and I just had to go ahead and pick up the pieces later. I found the whole process, especially seeing my leg for the last time before the operation, sickening and macabre. But in some ways the loss of my leg made me more determined to survive the cancer: I didn't want to have gone through all this for nothing.



2005 was without doubt the longest year of my life but finally I am starting to see light at the end of the tunnel. My three-month scan after finishing chemotherapy was clear and I am learning to walk again with a prosthetic limb. I have found I can ride a bike better than I can walk (although I fall off more often than I used to) and I have taken up horse riding again. I recently entered my first Riding for the Disabled competition – and won! Now I'm getting officially graded so I can compete nationally.



Best of all, this year I will be having my postponed wedding to the man who supported me through it all. We are lucky to be getting married in the absolute knowledge that we can make it through the tough times – because we already have.

BONE CANCER RESEARCH TRUST

The Bone Cancer Research Trust was formed in 2005 and has been registered as a charity. The Trust is an alliance of a number of established local charities and groups of family and friends of Primary Bone Cancer patients throughout Britain and Ireland. They share a common goal – to promote research into the causes and treatment of Primary Bone Cancer, and in particular of osteosarcoma and Ewing's sarcoma.

The main aim is to raise and distribute funds for research projects and programmes which address the causes, behaviour and treatment of bone sarcomas.

The Trust membership includes patients, medical practitioners and anyone interested in supporting research into these disabling and life-threatening diseases. The Trust is also looking to provide information, support and, in the longer term, counselling services for those suffering from primary bone cancer, and their families.



NEW DRUG TO TREAT GIST

A new treatment for GIST patients who have relapsed following treatment with imatinib (Glivec® from Novartis) has received a market licence in the USA and is expected to receive a licence for the UK and Europe this summer. Sutent® (also known as sunitinib) is manufactured by Pfizer and has shown effectiveness for a significant proportion of patients in this difficult condition, delaying the onset of disease in trials by an average time of about six months in those who respond.



Sutent is a targeted therapy which acts on the tumour cells to inhibit their ability to reproduce. It is taken as a capsule according to a schedule prescribed by the doctor. When licensed it will not automatically be available on the NHS and it is likely to be subject to a NICE review.

Clinical Trials for Sarcoma

The following are treatment trials currently available. There may also be trials available at individual hospitals, which local oncologists will know about.

A very good on-line trials database is at www.cancerhelp.org.uk/trials/trials/default.asp. If you want further information about trials there are also links on the Sarcoma UK website – www.sarcomauk.org/trials.htm

Where a trial is only held at one location a referral can usually be arranged. If you think a trial might be valuable for your treatment please talk to your consultant.

Trial Title	Description
EORTC 62991	Phase II pilot of moderate radiotherapy for inoperable aggressive fibromatosis (Desmoid tumours)
EURO-E.W.I.N.G. 99	Phase III pan-European treatment study for Ewing's sarcoma and Primitive Neuroectodermal Tumours (PNET)
EORTC 62012	Phase III Randomised trial of single agent doxorubicin versus doxorubicin plus ifosfamide in the first line treatment of advanced or metastatic soft tissue sarcoma.
Gemcitabine and docetaxel in leiomyosarcoma	A Phase II trial to assess the activity of gemcitabine and docetaxel as first line chemotherapy in patients with unresectable leiomyosarcoma. Only available in Royal Marsden & UCH, London.
EURAMOS 1	A Phase III randomized trial of the European & American Osteosarcoma Study Groups - strategies for resectable osteosarcoma based on response to pre-operative chemotherapy.
EORTC 62024	Phase III study of adjuvant Imatinib (Glivec) versus no further therapy after complete surgery for intermediate and high risk GIST.
VORTEX	Phase III randomized trial comparing two volumes of post-operative radiotherapy for adult patients with extremity soft tissue sarcoma.
EORTC 62027	Phase II trial of imatinib for the treatment of unresectable and/or metastatic dermatofibrosarcoma protuberans (DFSP) and giant cell fibroblastoma (GCF). Available at the Christie, Manchester.

RESEARCH UPDATE

Research into sarcoma, its causes and its cures, is being done in hospitals and scientific research centres scattered across the country. It is becoming more common for patients entering clinical trials to be asked to consent to samples from their tumours being used for research. All the projects below are relying on such samples.

We highlight here some of the programmes which are under way around the country.

Institute of Cancer Research – Sutton, Surrey

Professor Kathy Pritchard-Jones and her team are leading research in a number of childhood cancers. The tissue bank of the UK Childrens' Cancer Study Group (UKCCSG) is based at ICR. Two research fellows are working on rhabdomyosarcoma, one of them funded by the Chris Lucas Trust. This sarcoma also affects a small number of adults.

Professor Colin Cooper has been looking for new biological markers that predict clinical outcome in sarcoma, focussing on leiomyosarcoma. His team is examining the relationship between gene expression profile and the development of metastasis. Using cDNA microarray, the team has identified a gene expression signature associated with metastasis that allows prediction of the future development of metastases from primary tumours. The finding may aid the tailoring of therapy for individual sarcoma patients.

Manchester – Christie Hospital

Dr Catherine West is running a tissue bank in association with the Vortex trial of radiotherapy (see page 4). The aim is to analyse which patients are most likely to have a distant recurrence despite the impact of local radiotherapy. If making such predictions proves possible in the future more aggressive forms of treatment may be offered to patients with a proven risk.

Leeds – St James's Hospital

Dr Sue Burchill is working with tissue samples from patients who have had surgery and who are entered in the clinical trial EuroEwing99. She tells us a bit more about this project.

Biological Studies in EuroEWING99

The survival rate for children and young adults with tumours of the Ewing's sarcoma family (ESF) is poor, especially when there is metastatic disease at diagnosis. By individualising the treatment according to risk factors better overall survival might be achieved. It would mean those with small tumours and localised disease may benefit from less intensive treatment, and those with more aggressive tumours may be identified for more intense and/or novel therapeutic strategies. Unfortunately clinical methods do not accurately predict the disease course in all patients. Therefore there is an urgent need for new predictors to help assess risk for individual patients. In the trial EuroEWING99, two strategies are being investigated.

Strategy 1. Some patients with apparently localised disease at diagnosis rapidly develop metastasis from which they subsequently die. This suggests these patients have low-level metastatic disease at diagnosis that is not detected by current methods. A highly sensitive laboratory test called reverse transcriptase polymerase chain reaction, is being evaluated. It is capable of detecting one single cancer cell in a peripheral blood or bone marrow sample. This may lead to the introduction of more sensitive tests to detect metastatic disease, affecting future patient management and improving outcomes.

Strategy 2. Unfortunately in ESF there are no good biological markers that accurately define risk groups. Markers are features of the tumour that regulate how fast the tumour grows, disseminates, or responds to chemotherapy agents. However gene alterations may determine which patients do well and which do badly. Therefore the relationship between the tumour genotype at diagnosis and outcome is being evaluated. This may lead to improved methods of identifying those patients with the most aggressive disease and if these can be found it may be possible to identify potential targets for new therapies.

THE WRONG TIME IN HOSPITAL

Never get the wrong time when you spend nights in hospital. You may be looking at the clock at regular intervals to make time pass by, sure that you have a safe view, but there is always that possibility that you might slip up, especially in the middle of the night as you peer into the murky Dickensian light of the isolation room. Of course with me it may well have been the blood transfusion doing spectacular things with my body, as it slowly eked its way in or it could have been my raised temperature that had been lurking for days that was the result of my oncoming illusion.

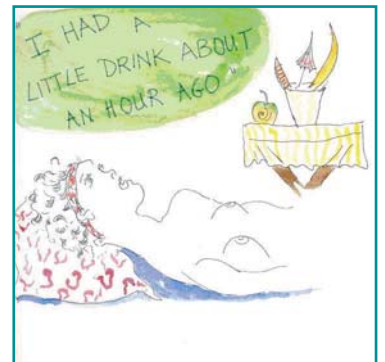
I had been having a difficult time that night lying there trying to sleep, with an uncomfortable body no matter which way I turned. But I had eventually got around to dreaming. The dreams were such blood thirsty adventures I could not possibly repeat though I will say I did not think that I could be such a savage. However, these dreams were eventually dispelled by a nurse slipping in to take observations and check on saline drips. And after she left I looked at the clock - 4 o'clock precisely, it said. Now that was alright, at least I had slept for a while and the promise of morning had already arrived because I was certain I could see a subtle light emanating from between the Venetian blinds.

With this optimistic thought in mind, I returned to dozing. The dreams I had from then on are repeatable, filled with crazy children's floats, families and cakes being sliced enthusiastically, a medieval pageantry combined with

a modern protest march all in darkness, if you understand what I mean! But as you can guess being on drips of one kind or another meant that the nurse had to enter again to check the line and this time take bloods too. The dreams came to a halt once more and I asked what the time was. She told me 4 o'clock, and I couldn't believe her! Or rather I did not wish to believe her, but there was the darkness pressing in on me and no ensemble of bird song nearby what so ever to tell me other wise.

"Are you sure?" I asked pathetically.

She smiled and nodded. So I got into foetal position once more, but this time not only were my pillows made of rough coals, but the little square window in the door projected a tiny beam of smug insistence from which I could not escape and a migraine was slowly but surely on its way. For comfort I placed my new fleecy dressing gown across the pillows with calm resignation tinged with renewed hope. Daylight had to come, and this madness would disappear. It was all a matter of more waiting.



This brief essay was written by Lynda Hopkins who lives near Henley-on-Thames. She was diagnosed with osteosarcoma on her left bicep and had 50% of the muscle removed plus some bone taken. This was followed by 2 months of chemotherapy. She has recovered well. She is left handed and has managed to regain full use of her arm to play piano and guitar and paint. One of her paintings recalling her stay in hospital is also printed.

RAISING AWARENESS OF SARCOMA

The Liddy Shriver Sarcoma Initiative is planning to raise public awareness of sarcoma worldwide. The event is called the “Team Sarcoma 2006 Bike Tour” and it is being held in Denmark on July 1-7, 2006. Tour organiser Bruce Shriver hopes that between 50-100 people will join the event, either as bikers or volunteers. Those booked so far come from Denmark, Ireland, the Netherlands, New Zealand, the UK, and several states within the US.

There are also “virtual Team Sarcomas” in many places worldwide for people who want to help raise awareness of sarcoma, but can't come to Denmark. People on virtual Team Sarcomas bike, run, walk, or swim on one or more of the days that the main team are biking in Denmark. Virtual Team Sarcoma groups in England, Poland, Portugal, Sweden, Ukraine, and in several states within the US.

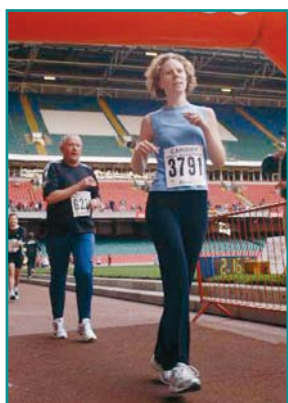
The Danish press intends to cover the bike tour in the newspaper and on TV and Scandinavian Airlines (SAS) will publish an article in its flight magazine, Scanorama, about sarcoma in June. Four million people read their flight magazine each month.



Participants in the Team Sarcoma 2003 bike ride

If you would like to take part, either in the bike ride in Denmark or as a ‘virtual’ participant contact Bruce Shriver at shriver@genesis2.com or contact Sarcoma UK and we will put you in touch. The Liddy Shriver Sarcoma Initiative’s homepage is at www.liddyshriversarcomainitiative.org

Sarcoma UK would like to express its thanks to those people who have made donations to our work, and who have supported sarcoma research efforts with our help.



Hanna Wilson

The Cardiff half-marathon was completed by Hanna Wilson and Matt Hopkins. Sharon Sanghera completed the Great North Run.

Lisa Colbeck has been fundraising to specifically support research on rhabdomyosarcoma. Graham White has formed the Gill White Trust and has initially raised over £2000 from a Quiz Night to support Professor Judson’s research at the Royal Marsden Hospital.

Sarcoma UK has been able to aggregate donations and make a grant to Dr Michael Leahy’s work with the Sarcoma Unit at the Christie Hospital in Manchester which will help increase the numbers of patients who can access clinical trials.



Matt Hopkins



Roger Wilson (Sarcoma UK) receives a cheque from Inspector Mike Milton (Calverton and Woodborough Pub Watch) and Angie Taylor (Calverton WMC Treasurer)

In March we received a generous donation from the Calverton & Woodborough Pub Watch scheme in Nottinghamshire from an evening hosted by Calverton Working Men’s Club.

Sarcoma UK uses donations to fund development of its information services and to promote patient interests in UK treatment centres. We also maintain a research fund which allows us to make small grants to support individual research projects and to stimulate research into sarcoma.

GETTING INFORMATION

Patients have very different needs regarding information. Sources of reliable information on sarcoma are improving all the time and on this page we try and give you a picture of what is available.

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

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.

General sarcoma information is available from Cancer BACUP. They have good free booklets on *Understanding Soft Tissue Sarcoma* and *Understanding Primary Bone Cancer*. There are also booklets on Radiotherapy and Chemotherapy. Your hospital information centre may have them, if not call Cancer BACUP (phone number below) and they will post them to you straight away.

If you have access to the Internet there is a wide range of information available from around the world but quality is variable. Some of the best sites are listed below.

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.

Contact telephone numbers and useful websites

CANCER HELP UK

Cancer Help is Cancer Research UK's website for patients. Excellent sarcoma material including "Questions for Your Doctors". Click the link to 'Specific Cancers' and 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 and other cancer related issues - all available free by phoning Bacup.

www.cancerbacup.org.uk/catalogue/sarcomas.htm

0808 800 1234 - freephone helpline

020 7696 9003 - for publications

MACMILLAN

Macmillan works to improve the quality of life for people living with cancer. It offers information and practical advice over the phone.

Macmillan also has information on local support groups.

www.macmillan.org.uk/

0808 808 2020 - freephone helpline

SARCOMA UK

Our website offers an introduction to all kinds of sarcoma and there are extensive links to a range of further Internet resources.

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

GIST SUPPORT UK

The UK's GIST support group with online information about treatments.

<http://www.gistsupport.co.uk>

BONE CANCER RESEARCH TRUST

BCRT is run by parents and patients to improve bone cancer research and support patients.

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

UK CHILDREN'S CANCER STUDY GROUP (UKCCSG)

UKCCSG co-ordinates the work of 22 specialised children's cancer centres across the UK. Website has useful range of advice leaflets.

www.ukccsg.org/public/resources/index.htm

CLINICAL TRIALS

If you are considering entering a clinical trial this site gives good general information about clinical trials.

<http://www.nelh.nhs.uk/clinicaltrials/>

Cancer Help also carries up to date information about the availability of clinical trials.

<http://www.cancerhelp.org.uk/trials/trials/default.asp>

INDUSTRY SITES

Novartis - Glivec®

Pharmamar - Yondelis®

Pfizer - Sutent®

Ariad - AP23573

www.novartisoncology.com/home.jsp

www.pharmamar.es/

www.pfizer.com/

www.ariad.com

SUPPORT GROUPS

There are sarcoma specific support groups at the following hospitals:

Royal Orthopaedic, Birmingham (ROHBTS)

Christie Hospital and Manchester Royal Infirmary, Manchester

City Hospital and Freeman Hospital, Newcastle

EMAIL SUPPORT GROUP

Provided by Sarcoma UK

<http://www.mailtalk.ac.uk/lists/sarcoma.html>

Be cautious when using the Internet 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 cannot be specific to a particular patient.