Gynaecological Sarcoma: The Hidden Cancer
About Sarcoma UK

Sarcoma UK is the only charity in the UK focusing on all types of sarcoma.

Our mission
To increase knowledge and awareness of sarcoma through ground-breaking programmes that inspire involvement and transform the landscape for everyone affected by sarcoma.

- We raise sarcoma awareness to initiate change and improve standards of treatment and care
- We seek answers through research
- We provide support & information for the sarcoma community

Acknowledgements
We would like to thank

- the 55 women and family members who completed the survey;
- sarcoma clinicians and clinical nurse specialists in sarcoma centres around the country who distributed the questionnaire to their patient;
- sarcoma clinicians who advised on the content of the report;
- women with gynaecological sarcomas who commented on the content of the report.

Production
Survey design: Claire Kelleher and Lindsey Bennister
Data analysis and comment: Lindsey Bennister, Claire Kelleher and Roger Wilson CBE
Report author: Lindsey Bennister
Design: INQ Design Ltd 020 7737 5775
**Introduction**

_Gynaecological Sarcoma: The Hidden Cancer_ draws on the personal experiences of women with gynaecological sarcoma who took part in a Sarcoma UK survey in 2014. It contains both quantitative and qualitative data.

The report is part of the charity’s *Sarcoma Voices* programme, which gathers the experiences of people affected by sarcoma in order to influence and improve sarcoma services, treatment and care.
At Sarcoma UK, we have become increasingly aware of the devastating impact of gynaecological sarcomas through our contact with women and their families and from reports given to us by specialist sarcoma doctors. There is frustration because the problems women face mean that many cannot access the best treatment and in some cases do not receive appropriate treatment.

This survey tells us what is actually happening. It shows us a health service where women have to fight to be taken seriously right from the point when they first present with symptoms. A health service which often delays referral to a specialist sarcoma service until disease is too advanced for more than palliative treatment.

For the 280 women and their families affected by gynaecological sarcomas each year, this is simply not good enough.

The survey also tells us that there is good practice happening in parts of the health service. In some areas there are exemplary clinical pathways which are diligently observed. Some women have had textbook referrals and treatment. This gives us hope. We need to build on this.

This report concludes with strong recommendations that may extend or save lives and will certainly improve the experiences of women and their families affected by gynaecological sarcomas. More lives could be saved through relatively small and simple changes.

Our heartfelt thanks go to all the women who took the time to share their experiences with us. We would also like to thank anyone who completed a questionnaire on behalf of someone close to them who sadly is no longer with us because of this devastating disease.

It is our hope that this report increases awareness and understanding of gynaecological sarcomas so that it no longer a hidden cancer.

Lindsey Bennister  Chief Executive
Sarcoma UK is the only charity in the UK focusing on all types of sarcoma. We provide support and information to women and their families affected by gynaecological sarcomas.

This survey is the first time that the experiences of women with gynaecological sarcomas throughout the UK have been collected and recorded.

Gynaecological sarcomas make up 13% of all sarcomas; and 3-4% of all gynaecological cancers are sarcoma. Approximately 280 women are diagnosed with a gynaecological sarcoma each year.

Survival rates for gynaecological sarcomas in the UK are poor and haven’t improved significantly in the last two decades, especially compared to other countries. For example, five year relative survival for the most common gynaecological sarcoma – uterine leiomyosarcoma – is only 37%. There is great potential to improve this situation through better care and treatment and a more consistent approach to the management of gynaecological sarcomas.

NICE Improving Outcomes Guidance for Sarcoma currently recommends a ‘shared care’ approach for women with gynaecological sarcomas. Evidence points to limited progress with the establishment of effective clinical pathways, and little consensus between gynaecological oncology and sarcoma specialists.

There is evidence of good practice in some parts of the country: textbook referrals by GPs to appropriate secondary care; exemplary clinical pathways; and effective shared care where gynaecological oncology and sarcoma teams work together in the best interests of their patients.
Executive Summary

Key Findings

1. Reaching a diagnosis
   - Women experience symptoms for many months and even years before being referred for investigations. Younger women in particular visit their GP numerous times before being referred.
   - There are red flag symptoms that should raise suspicion of gynaecological sarcoma. These include: persistent and sometimes severe bleeding that is getting worse; fibroids that are increasing in size (particularly after menopause); persistent abdominal pain that is getting worse; and lumps that are increasing in size.
   - Most women are referred initially to a gynaecologist.
   - There are inconsistencies in the use of imaging in the diagnostic process, with a quarter of women reporting no imaging at all prior to surgery.
   - There is a major problem with the diagnostic confirmation of sarcoma in terms of both accuracy and the length of time it takes. Most women waited weeks; some waited over two months; and two women waited eight and 12 months for their diagnosis post-surgery.

2. Accessing best care and treatment
   - A large proportion of women are not referred to sarcoma specialist services following a diagnosis of sarcoma, a recurrence, or on diagnosis of metastases.
   - Many women only reach sarcoma specialist services when they themselves insist on it, or as a last resort when treatment options have come to an end.
   - Many women are not receiving appropriate and effective treatments for gynaecological sarcomas. This survey identifies cases where treatments are given with no evidence of efficacy, for example adjuvant therapies that are not standard of care for sarcoma. As a result, women experience significant side effects that impact on quality of life, or may result in an increased risk of a recurrence.

3. Support and information
   - The majority of women do not see a clinical nurse specialist during their diagnosis and treatment, denying them access to vital emotional support and information about their treatment options and care.
Executive Summary

Recommendations

1. A consensus position and guidelines on the management of gynaecological sarcomas should be agreed between gynaecological oncology specialists and sarcoma specialists via the professional bodies.

2. A symptoms index for gynaecological sarcoma should be developed and adopted.

3. NICE Improving Outcomes Guidance for Sarcoma should be updated to include new and clear guidance for the management of gynaecological sarcomas.

4. Effective clinical pathways for the referral, diagnosis and care of women with gynaecological sarcomas should be in place across the NHS to facilitate the shared care approach to management. These should build on existing good practice that is in place in some regions.

5. Sarcoma specialist pathologists should be involved in confirming any diagnoses of gynaecological sarcoma.

6. All gynaecological sarcomas should be tested for hormone receptors as part of their diagnosis.

7. Sarcoma Multi-Disciplinary Teams should include members with appropriate expertise in gynaecological sarcomas. If this is not available, patients should be referred to other sarcoma services with such expertise.

8. Every woman diagnosed with a gynaecological sarcoma should have access to a clinical nurse specialist during every stage of their treatment and care.

9. Sarcoma specialised services should remain within NHS England specialised commissioning.
The female reproductive system

- Fallopian tube
- Ovary
- Endometrium
- Uterus
- Cervix
- Vagina
Gynaecological Sarcoma: The Hidden Cancer

1. Gynaecological sarcomas

Sarcomas are rare cancers that develop in the connective tissues: muscle, bones, nerves, cartilage, tendons, blood vessels and the fatty and fibrous tissues.

Gynaecological sarcomas, sometimes shortened to gynae sarcomas, occur in the female reproductive system: the uterus (womb), ovaries, vagina, vulva and fallopian tubes. They can affect women of any age.

Three to four per cent of all gynaecological cancers are sarcomas and they make up 13% of all sarcomas. Approximately 280 gynaecological sarcomas are diagnosed annually.

Most gynaecological sarcomas (85%) occur in the uterus (womb) and 7% occur in the ovaries. The remainder occur less commonly in the vagina, vulva, fallopian tubes and other areas of the female reproductive system.

The main types of gynaecological sarcomas:

- 52% are leiomyosarcoma – a cancer of the smooth or involuntary muscle, mostly but not always occurring in the uterus
- 24% are endometrial stromal tumours – a cancer of the connective tissue in the uterus
- 16% are sarcoma not otherwise specified (NOS) and include undifferentiated endometrial sarcoma (UES)

The remaining 12% are mostly made up of:

- Rhabdomyosarcoma – a cancer of the skeletal muscle, mostly occurring in the uterus and vagina
- Fibrosarcoma – a cancer of the fibrous tissues, mostly occurring in the ovaries
- Liposarcoma – a cancer of the fatty tissues, mostly occurring in the ovaries, vulva and uterus

Background

Malignant Mixed Mullerian Tumour or carcinosarcoma is a mixture of carcinoma and sarcoma, where the carcinoma component is the dominant feature. These types of cancer are not treated as a sarcoma but treated in a way that is similar to other cancers (carcinomas) of the ovaries and uterus, by gynaecological oncologists.

1. Text from Understanding gynaecological sarcomas. Sarcoma UK. November 2014
**Background**

Whilst more women are surviving gynaecological sarcoma, there have been only slight improvements in survival in the past 25 years. Five year survival rates still remain below 50%\(^2\) and little has changed in the structures or clinical patterns of treatment to bring about significant improvements in the future. For women diagnosed with uterine leiomyosarcoma, the most common gynaecological sarcoma, the situation is much poorer with only 37% of women surviving for five years. This is worse than in other countries, for example Norway, where uterine leiomyosarcoma survival is somewhat higher at 48%.\(^2\) Women diagnosed with the second most common gynaecological sarcoma – uterine endometrial stromal sarcoma – fare better with a survival rate of 68% at five years.

**2. Management of gynaecological sarcomas**

Throughout the health services of the UK, gynaecological sarcomas pose a particular challenge. Inconsistencies and lack of clarity in management are likely to be strong contributory factors to the low survival rates for these sarcomas.

NICE Improving Outcomes Guidance for Sarcoma (2006)\(^3\) recommends a 'shared care' approach for women with gynaecological sarcoma. This recognises that initial referral is most likely (and appropriately) to a gynaecological specialist, followed by liaison with the sarcoma Multi-Disciplinary Team (MDT), usually once sarcoma is diagnosed. Shared care is of particular value in the pathological confirmation of sarcoma and the consideration of adjuvant therapy following surgery and diagnosis. Monitoring and treatment for metastases is best delivered in line with sarcoma protocols, and the provision of chemotherapy is best delivered under the supervision of sarcoma units. In advanced disease, surgery for lung metastases is frequently carried out for soft tissue sarcomas generally, but much less for gynaecological sarcomas. This may reflect that many women with gynaecological sarcomas are not seeing sarcoma specialists.

However, progress with the establishment of effective pathways for shared care has been slow. In addition, we recognise that some sarcoma MDT’s are not always equipped to deliver advice on gynaecological sarcomas due to lack of experience within the MDT.

**3. Sarcoma UK’s position on shared care**

If gynaecological sarcoma is diagnosed before surgery, following investigations and tests, treatment should be managed jointly by a sarcoma MDT and a gynaecological oncology (cancer) MDT. Surgery for gynaecological sarcoma should ideally be carried out by specialist surgeons with experience of gynaecological sarcoma. This is because gynaecological sarcomas are often poorly differentiated and much of the treatment is based on expertise gained during the management of patients with soft tissue sarcomas.

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\(^3\) NICE Improving Outcomes Guidance for Sarcoma. 2006
out by a surgeon with specialist expertise in gynaecological cancers, working together with sarcoma specialists to ensure the right treatment is given.

If gynaecological sarcoma is diagnosed after surgery, a referral should be made immediately to a sarcoma MDT with experience in gynaecological sarcoma who will review the case, manage ongoing care, and decide on the best treatment options available in the future.

4. Survey methodology

Between February 2014 and August 2014, Sarcoma UK developed and distributed a survey to women affected by gynaecological sarcoma.

The survey asked 47 questions – see Appendix B.

The survey was available online and in paper format. It was promoted widely

- on Sarcoma UK’s website;
- to an online gynaecological sarcoma support group;
- by sarcoma clinical nurse specialists and consultants in eight sarcoma treatment centres;
- to the network of 15 regional sarcoma support groups;
- through Sarcoma UK’s social media.

We note the different health services in Scotland and Northern Ireland and acknowledge that the recommendations in this report relate mainly to NHS England services. However, we believe that the findings of this survey will be of interest to patients and clinicians throughout the devolved nations in relation to developing best practice and improving the experiences of women with gynaecological sarcoma.

5. Responses

- 55 questionnaires were completed, including three questionnaires that had been completed on behalf of a family member/partner who had passed away.
- 61% of respondents had been diagnosed within the last three years.
- 27% of respondents had been diagnosed in the previous year (2013).

We recognise that this is a relatively small sample and that there may be some bias in the results as this was a self-selected group. However, as a proportion of the overall incidence of sarcoma, it is roughly equivalent to an 8,500 patient study in lung cancer. This is both a reflection on the rarity of sarcoma and on the validity of this sample of patient experience.

“27% of respondents to our survey had been diagnosed in the previous year.”
Results

1. Age at diagnosis
This ranged from 22 to 69 years and the median age was 48. There were no respondents aged 70 or above.

This data indicate that women are diagnosed with gynaecological sarcomas at all ages and stages in their life. Just under 10% of respondents were in their 20’s, and 16% were under 40 years old.

2. Symptoms
The open text responses indicate a common set of symptoms that women experienced prior to diagnosis. These are:

- persistent and sometimes severe bleeding that is getting worse
- fibroids that are increasing in size (particularly after menopause)
- persistent abdominal pain that is getting worse
- lumps that are increasing in size

The duration of symptoms prior to diagnosis varied enormously. Two patients had their tumour found as a direct result of other procedures and attributed no symptoms to the cancer. Several had complained of relevant symptoms to their GP for four to five years. The range of symptoms duration was 0 to 60 months.

Women who experienced the longest duration of symptoms (over two years) had a
median age of 53. The median time of symptoms for those under the age of 40 was 12 months, more than 50% higher than for the whole group of respondents.

3. Referral for investigations
We asked respondents how many GP visits they had made before referral to a specialist for investigations.

More than half of respondents only had one GP visit before referral to a specialist for investigation. However, for the other half it was a very different picture with several respondents stating that visits were so numerous that they could not recall the number. Where a number was given, the average was two visits.

The average age of those who only required one visit to the GP before referral was 50. Of the under-40 age group, just under half had four or more GP visits, with one respondent in her 20s visiting the GP 20 times.

“I went to the GP with a lump in my labium maiora (part of the vulva). I was examined although it didn’t feel like the lump was examined properly. I was told it was probably to do with shaving the area and not to shave for a while. When I suggested that I should come back in two weeks if it hadn’t improved, I was greeted with a simple shrug of the shoulders.”

One respondent insisted on a referral to a specialist and was told by the GP that she would be wasting the consultant’s time, while another reported her symptoms to the consultant treating her for breast cancer and was rapidly referred on.

Eighty two per cent of respondents were referred by their GP via the NHS; the rest were referred for investigations through the private healthcare route.

Eighty two per cent of respondents were referred to a gynaecologist while 18% were referred to a gynaecological oncologist, suggesting that there was some suspicion of cancer in the mind of the referring doctor. This may be the positive outcome of national awareness and education programmes around
gynaecological cancers such as ovarian cancer. None of the respondents were referred to a sarcoma specialist and this was not unexpected.

**Suspicion of cancer/sarcoma**
We asked women whether they were told that cancer, or a sarcoma, was suspected prior to surgery.

Just over half of respondents (53%) went into surgery having no idea their problem might be cancerous. It is also likely that their gynaecologist also didn’t suspect cancer.

Of the other half of respondents, the majority (33%) were warned that a cancer might be found and 14% were informed that a sarcoma was suspected.

### 4. Diagnosis of sarcoma

**Initial diagnosis**
The initial diagnosis received by respondents that prompted further investigations and treatment varied enormously. Forty per cent of respondents were told that their symptoms were due to fibroids. Other initial diagnoses included

- problems caused by contraceptive devices;
- general harmless mass;
- cyst/polyp;
- ovarian cancer.

**Use of imaging**
We asked women about the imaging that had been done prior to surgery.

Twenty six per cent of the respondents had no imaging prior to surgery.

Several respondents had more than one type of scan. Around one-third of respondents had each of these scans.

*Several respondents had more than one type of scan*
Results

**When was a diagnosis of sarcoma made?**

Sarcoma was confirmed prior to surgery for only 16% of respondents, following imaging (10%) or a biopsy (6%).

The majority of respondents (84%) were told that they had a diagnosis of sarcoma following surgery.

**How long did it take to receive a diagnosis of sarcoma?**

The median number of weeks post-surgery that respondents waited to receive a diagnosis of sarcoma was four. However, the range was very wide. Two respondents received a diagnosis immediately, whilst seven respondents (13%) waited two months or more including two respondents who waited eight and 12 months respectively.

**Metastases at diagnosis**

Sixteen per cent of respondents had sarcoma that had already spread to another part of their body (metastatic disease) at diagnosis. Two women were referred initially for investigation into what turned out to be secondary cancers, where the primary gynaecological sarcoma was found later on.

Neither of these received surgery for their sarcoma.

Only one respondent with metastases at diagnosis was diagnosed with sarcoma pre-surgery, following a biopsy, CT and MRI scan. She was treated by a gynaecological oncology team receiving radiotherapy pre-surgery followed by a total abdominal hysterectomy and bilateral salpingo-oophorectomy (ovaries removed).

There was no consistency in the type of imaging used with women who had metastases at diagnosis. One respondent had no imaging at all and only a biopsy. Three respondents had a CT, MRI, and biopsy. Others had ultrasound and x-ray imaging in various combinations. There was no difference in NHS or private imaging practice.
5. Treatment

**Initial treatment**
The majority of respondents (69%) had open surgery (laparotomy) with a total abdominal hysterectomy and bilateral salpingo-oophorectomy.

Other surgical treatments included:
- open surgery total abdominal hysterectomy (ovaries not removed) – 8%
- vaginal hysterectomy – 8%
- myomectomy (surgery to remove fibroid but preserve uterus) – 8%
- laparoscopic procedure – 2%

Three respondents did not have an option of surgical treatment and were treated with chemotherapy. Two of these were treated by a gynaecological oncologist (and were not referred to a sarcoma specialist).

“I was treated for eight years by a very good oncologist but he did not specialise in sarcoma. I did not have frequent CT scans and that’s why secondaries were not spotted.”

**Adjuvant treatment**
Over half of all respondents had adjuvant therapy (additional treatment options following initial surgery).

The most popular type of adjuvant therapy was radiotherapy. Over a third of respondents received some form of radiotherapy in the pelvic area following surgery, either to the original tumour site or more regionally in the pelvis.
Twenty per cent of respondents received adjuvant chemotherapy. The types of chemotherapy used varied considerably and included:

- Docetaxol + gemcitabine
- Docetaxol + carboplatin
- Doxorubicin
- Doxorubicin + ifosfamide
- Doxorubicin + cisplatin
- VIDE (vincristine, ifosfamide, doxorubicin, etoposide)

Of those respondents who received adjuvant therapy, 51% did not have their treatment overseen by a sarcoma specialist.

Recurrence rates

Just under half (48%) of respondents had a recurrence of their sarcoma; 70% of these had metastases in the lungs, liver and other sites.

6. Reaching sarcoma specialist services

No respondents were referred to a sarcoma specialist for investigation prior to diagnosis.

Only half of respondents were referred to a sarcoma specialist following their diagnosis of sarcoma.

The other half of respondents remained under the care of a gynaecological oncology team or a gynaecologist, despite having a confirmed diagnosis of sarcoma. Of these, only one respondent was told that a sarcoma specialist had been consulted to agree on a care pathway.
Fifty-one per cent of those respondents who had adjuvant treatment did not have their treatment overseen by a sarcoma specialist.

Seventy-two per cent of respondents who had a recurrence of their sarcoma were not referred to a sarcoma specialist at that stage.

7. Hormone responsive sarcomas
Some gynaecological sarcomas express hormone receptors (oestrogen and progesterone) on the tumour cell surface; the hormones can bind to the receptors on the tumour cells and may drive the cancer and stimulate it to grow. Half of leiomyosarcomas in the uterus express hormone receptors on the tumour cell surface, and endometrial stromal sarcomas almost always express hormone receptors. There is evidence that patients with endometrial stromal sarcomas respond to hormonal treatments that block the effects of oestrogen and progesterone on the tumour cells. There is more limited evidence that selected patients with uterine leiomyosarcoma may also benefit from hormonal treatment.

We asked women whether their tumour had been hormone tested:
- 76% of respondents said their tumours had been hormone tested
- 62% was following initial surgery by the surgical team
- 38% was following a recurrence of sarcoma

Of the respondents who had their tumours hormone tested, 76% had positive results, ie they had tumours that expressed oestrogen and progesterone receptors. Ideally all gynaecological sarcomas should be tested for oestrogen and progesterone receptor expression, as hormonal treatment may be indicated for some patients.

8. Support
Fifty-one per cent of respondents did not receive any support from a clinical nurse specialist.

Only 8% of respondents received support from a sarcoma clinical nurse specialist.
We have highlighted three areas of major concern:

1. Reaching a diagnosis
Our results clearly indicate that receiving a prompt referral and diagnosis of gynaecological sarcoma is a significant problem. Many women experience symptoms that should raise concern and lead to a prompt referral for investigations.

a. Symptoms
The free text element of the questionnaire offers insights into the symptoms women experienced, enabling us to draw some conclusions about the red flag symptoms for gynaecological sarcoma that could be used to guide further investigations. The majority of respondents experienced one or more of these symptoms: persistent and sometimes severe bleeding that was getting worse; fibroids that had increased in size (particularly after menopause); persistent abdominal pain that was getting worse; and lumps that were increasing in size. These symptoms match those identified in (as yet unpublished) data from a leading sarcoma centre. Forty percent of respondents were told that their symptoms were due to fibroids. It is vital that a way of distinguishing between benign fibroids and malignant cancer is identified. These red flag symptoms may be a starting point for discussion.

b. Prompt referral
Age appears to be a key factor influencing how quickly women are referred for investigations. In our survey, women in their 50’s had symptoms for the longest duration (over two years) but were on average referred for investigation after their first visit to their GP. This contrasts sharply with the experiences of younger women (under 40) who also experienced symptoms for a long time – the median time was 12 months which is more than 50% higher than for the total group of respondents. However, younger women visited their GP many more times before being referred for investigation, with half of this group visiting their GP four times or more. This may be explained by the poor level of GP suspicion of cancer in the younger age range.

Discussion

"These red flag symptoms may be a starting point for discussion."

"Age appears to be a key factor influencing how quickly women are referred for investigations."
Many respondents described the struggle they had to get their worries about symptoms taken seriously by their GP. A number of women indicated that they had to be forceful and insist on a referral, often against their GP’s advice. One woman was told she would be wasting the consultant’s time. The qualitative element of the survey provides more insights.

Data from this survey also indicate a large variation in the types of investigations women received before treatment, and little consistency in approach. Forty per cent of respondents were told that their symptoms were due to fibroids and this clearly points to a lack of suspicion of cancer (and sarcoma) by gynaecologists.

c. Imaging

A quarter of women reported having no imaging at all prior to surgery. Our data show no pattern or consistency in the types of scans women received.

We recognise that diagnosing a gynaecological sarcoma which has an incidence of 1:350 incidence is demanding. However, there is currently an absence of any clear consensus on the optimal imaging of gynaecological sarcoma (in particular in the uterus). This is reflected in inconsistent practice and lack of guidelines for radiologists on how to interpret images. Given the proven value of CT/MRI in diagnosing other forms of sarcoma, this is a major problem.

d. Prompt diagnosis on discovery of cancer

Most women were diagnosed post-surgery, following analysis of tissue by a pathologist. The length of time that respondents waited for a diagnosis varied considerably but their experiences point to a major problem with the diagnostic confirmation of sarcoma. It is unsatisfactory that 13% of respondents waited two months or more for a diagnosis of sarcoma. For two respondents to have waited eight and 12 months is unacceptable and raises serious questions about the diagnostic pathways of those health service providers involved.

The NICE Improving Outcomes Guidance for Sarcoma recommends that a sarcoma diagnosis should be made by a specialist pathologist with expertise in sarcoma. The experiences of our respondents clearly indicates the importance of involving sarcoma specialists (and the sarcoma Multi-Disciplinary Team [MDT]) at the earliest possible opportunity.
2. Accessing best care and treatment

This survey provides evidence of problems accessing best care and treatment for gynaecological sarcoma.

a. Reaching sarcoma specialist services

The NICE Improving Outcomes Guidance for Sarcoma states that the care of patients with soft tissue sarcomas that require shared management should be managed by the appropriate site-specific Multi-Disciplinary Team (MDT) in conjunction with a sarcoma MDT. This means that the site specific MDT has primary responsibility to liaise with the sarcoma MDT to discuss the management of each patient. However, our survey clearly indicates that this is not happening in practice. Even following a confirmed diagnosis of sarcoma, a recurrence, or a diagnosis of metastases, a significant number of women with gynaecological sarcomas are still not referred to sarcoma services. It appears there is a resistance on the part of some gynaecologists and gynaecological oncologists to refer to a sarcoma team. This resistance can also be surmised from the comments of women in our survey. One respondent described having to “fight to be heard” when she asked for her case to be considered by a sarcoma specialist. Another described the effort required “to push to get seen by the right people” and having “a terrible argument” with their gynaecologist.

Our results also indicate that many women find that they are only referred to a sarcoma specialist team when the (non-sarcoma) professionals are unable to identify any further treatment options, often as a last resort. This is a frustrating and unsatisfactory situation as women are not reaching sarcoma services at a point where they have optimum treatment options available to them. The personal upset this causes to women and their families is clearly illustrated in the qualitative section of this report.

This all points to a major failure in the implementation of the NICE Improving Outcomes Guidance for Sarcoma in relation to gynaecological sarcoma. It may in part be a result of limited understanding among local commissioners about the requirement for shared care between site-specific and sarcoma Multi-Disciplinary Teams. It may be explained by the stronger influence of gynaecologists and gynaecological oncologists locally compared to sarcoma clinicians and the lack of awareness of sarcoma. However, we know that improvements can be made. There are good examples of best practice where clear local
pathways are in place, providing effective shared care arrangements. The qualitative section of this survey makes reference to this.

**b. Receiving best treatment for sarcoma**

Over half of our respondents entered into treatment (usually surgery) with no suspicion that they may have cancer. Only a handful had been told about sarcoma before treatment started. Our survey identifies issues with the treatment women receive, particularly post-surgery, and the long-term impact this may have.

Our data indicate that 55% of respondents had adjuvant therapy following surgery, with over a third of respondents receiving adjuvant radiotherapy to the pelvic area. Twenty per cent of respondents received adjuvant chemotherapy. The explanation given to patients about the value of adjuvant therapy was to reduce the risk of the spread of cancerous cells post-surgery.

Our data show that half of respondents who received adjuvant therapy were not under the care of a sarcoma specialist. We question whether some of these treatments are appropriate for sarcoma and point to recent published papers\(^1\) that question the value of adjuvant radiotherapy for gynaecological sarcoma. Data also indicate that some women were undergoing adjuvant chemotherapy for gynaecological sarcoma when not under the care of a sarcoma specialist team. The wide variety of chemotherapy regimens cited by respondents points to a lack of understanding of effective treatments for gynaecological sarcoma by non-sarcoma clinicians, and a lack of an evidence-based approach to patient management. As a result, women may experience unnecessary and distressing treatments which are not effective for their type of sarcoma.

The decision about use of adjuvant therapies should be made by sarcoma specialists who are best placed to interpret the limited data available and to select women at higher risk of recurrence who may benefit from this type of therapy. Our data point to widespread use of adjuvant therapies rather than selective use for those women who may benefit.

**3. Support and information**

Emotional support and access to information is vital for women undergoing treatment for gynaecological sarcomas. The National Cancer Patient Experience Survey (2014) highlights the value of the clinical nurse specialist...
in providing support and information for cancer patients. It is a very unsatisfactory position that half of our respondents did not see a clinical nurse specialist at all during their diagnosis or treatment. This further indicates the lack of clarity around clinical pathways and where responsibility for the care of gynaecological sarcoma patients lies.

Access to support and information is particularly important for those women whose sarcoma was diagnosed following routine surgery and were not anticipating a cancer diagnosis. It is also vital for younger women who may want to consider fertility options pre- and post-treatment.
Recommendations

1. Sarcoma UK calls on the professional bodies representing gynaecological specialists, gynaecological oncologists, and sarcoma specialists to develop, agree and implement a **consensus position and guidelines on the management of gynaecological sarcoma patients**. This should cover diagnosis, referral and treatments. It should also set out a clear approach to imaging, particularly where fibroids are present, which can be adopted as standard across gynaecological practice.

2. A symptoms index for gynaecological sarcomas should be developed and adopted by professional bodies and promoted widely to GPs and gynaecologists to guide further investigations. **This should be based on a consensus of red flag symptoms experienced by patients.**

3. **NICE Improving Outcomes Guidance for Sarcoma should be updated** to include guidance for the management of gynaecological sarcomas, ideally based on the consensus position/guidelines developed by the professional bodies.

4. **Effective clinical pathways for the referral, diagnosis and care of women with gynaecological sarcomas should be in place across the NHS**, to fully establish the shared care approach to management. These should build on existing good practice that is in place in some regions. Pathways should be clearly communicated to GPs to assist them to refer promptly to appropriate specialists.

5. Sarcoma specialist pathologists should be involved in confirming a diagnosis of gynaecological sarcoma, with clear clinical pathways in place to enable this to happen promptly so that effective treatment can begin as early as possible.
6. **All women** who have a confirmed diagnosis of leiomyosarcoma or endometrial stromal sarcoma **should have their tumours tested** for oestrogen and progesterone receptor expression in order to identify patients who might benefit from hormonal treatments.

7. **Sarcoma Multi-Disciplinary Teams should include members with appropriate expertise on gynaecological sarcomas.** If this is not available within their service, prompt referral should be made to another sarcoma centre which has relevant experience.

8. **Every woman** diagnosed with a gynaecological sarcoma **should have access to a clinical nurse specialist** during every stage of their treatment and care, to provide support and information.

9. **Sarcoma specialised services must remain within NHS England specialised commissioning,** ideally as a nationally commissioned service. Local commissioning or co-commissioning of sarcoma services is unlikely to address the issues raised in this report or bring about solutions to improve outcomes for women with gynaecological sarcomas.
Appendix A: Qualitative data

These personal experiences are taken from the open text section of the survey questionnaire.

“Diagnosis to treatment was very quick and I had full support of a specialist team involving a specialist sarcoma team and gynaecology oncology team. Macmillan nurse support and local hospice was invaluable. Some groinal lymph nodes were left (we think three) and I suffer from lymphoedema following surgery but support is available for this via the sarcoma centre and hospice.”

“Two pathologists gave two different opinions. One positive, the other negative.”

“I was diagnosed with a bartholin cist in my vulva in February 2007. The gynae doctor said to leave alone and that it would never become cancerous. It began to grow and in two years had gone from the size of a small marble to the size of a large free range egg. I had it removed as it was getting in the way. Two weeks later I had a phone call – I needed to see the doctor urgently. The lump had been analysed and it was leiomyosarcoma.”

“My initial diagnosis was as the result of a tumour on my spine which turned out to be a leiomyosarcoma. It was then discovered that the primary was in my womb and that I had lung mets. As soon as the spine tumour was identified, I was referred to a sarcoma specialist. My care is currently under sarcoma specialist unit, neurosurgeon and gynaecology/oncology surgeon. I knew I had fibroids but had no idea they could turn into cancer. I had no symptoms until the secondary tumours.”

“The tumour was inside a large fibroid which was not detected with an ultrasound. If it had been, there may have been a different story.”
“My first symptom was bleeding – a hysteroscopy/biopsy gave initial diagnosis. Radiotherapy was needed to reduce the sarcoma to be operable. I was told the hysterectomy was a success with all traces of sarcoma removed. I continued as a gynaecology patient which I thought odd – wombs don’t come back but sarcomas do! Mine returned with the first symptom being severe abdominal pain. I had a scan and an operation to remove mass. THEN I was referred to a sarcoma specialist.”

“I asked the oncologist if sarcoma specialists were being consulted by him but I haven’t received a clear answer.”

“In a way, I deeply regret having radiotherapy as years later I have bowel problems (not too serious) and pelvic fractures (very serious). I wish I had more advice about the radiotherapy at the time, including long-term effects. I also feel I should have been monitored for bone density, at least. I think everyone should be monitored in this way. I cannot now find a consultant who specialises in fractures due to radiotherapy. This is strange because I am told that fractures after radiotherapy are rather common.”

“My mum had a terrible argument to get to see a sarcoma specialist. Only after the second recurrence and after pushing did she see a sarcoma specialist. By then it had spread everywhere. My mum wrote a diary about her experience and how hard she had to push to get seen by the right people.”

“My sarcoma was diagnosed in the private system. I pursued a second opinion through my GP when I realised the rarity of sarcoma and the need for specialist knowledge of the disease. My treatment is now with the NHS and I still feel aggrieved that my initial gynaecologist and oncologist NEVER suggested specialist intervention. Indeed, if I had not been so pro-active and followed other sarcoma patients’ advice, I would be in a much worse situation than I am today, thankfully!”

“Once I was referred to the sarcoma specialist my care and treatment went very well.”
“My oncologist advised me to have chemotherapy but also told me that my cancer was rare and aggressive and that no one had survived longer than two years. In my case, it could be as little as three months. I feel fit and well. We country dance three times per week, maintain our garden and regularly walk up to eight miles quite energetically (10 if we get lost!).”

“I feel that I have had to fight to be heard. If I hadn’t insisted on a CT scan after 10 months, then I don’t know how long would have been left. It was this scan that detected lung tumours.”

“There was controversy over whether I should have radiotherapy. I wish I hadn’t had it as I now have (8 years later) a fractured pelvis. This happened 18 months ago and it is still not healed and has caused painful sciatica and apparently stenosis.”

“It was frightening to have a nurse and oncologist shrugging their shoulders when you ask questions. When faced with a rare cancer, the very least they can do is refer to a specialist centre. I went back to my GP and requested a second opinion, however I was made to feel that I should never have asked for this and was wasting time.”
Appendix B: Survey questionnaire

Between February 2014 and August 2014, Sarcoma UK developed and distributed a survey to women affected by gynaecological sarcoma.

The survey asked 47 questions.

The survey was available online and in paper format. It was promoted widely

- on Sarcoma UK's website;
- to an online gynaecological sarcoma support group;
- by sarcoma clinical nurse specialists and consultants in eight sarcoma treatment centres;
- to the network of 15 regional sarcoma support groups;
- through Sarcoma UK’s social media.
Sarcoma UK is conducting a survey to gather women’s experiences of gynaecological sarcoma. We hope to use this information to highlight any issues or gaps in care so we can move towards improving the diagnosis procedures and treatment options available to women.

1. Date of diagnosis: 
2. Age at diagnosis: 
3. How long did you have symptoms before getting a diagnosis? 
4. How many GP visits did you make before you were referred to a specialist? 
5. Who were you referred to? 
   - Gynaecologist
   - Sarcoma specialist
   - Gynaecological oncologist
   - Other (please specify) 
6. Did your GP suggest any other treatment before specialist referral? 
   - Yes
   - No 
7. If yes, please give details, including if you proceeded with this treatment 
8. Was your GP referral NHS or privately routed? 
   - NHS
   - Privately routed 
9. What imaging, if any, was done by your GP before referral to a specialist? 
   - X-ray
   - CT scan
   - Ultrasound
   - MRI scan
   - None 
10. What imaging, if any, was done by your specialist prior to surgery? 
   - X-ray
   - Ultrasound
   - CT scan
   - MRI scan
   - None 
11. Did you have a biopsy (taking and testing a sample of tissue) prior to surgery? 
   - Yes
   - No 
12. What was the initial diagnosis and proposed action? 
13. Were you given a choice between different kinds of surgery? 
   - Yes
   - No 
14. If yes, were you given time to make a decision and written information to help make the decision? Please list any treatment or surgery options offered to you.
### Gynaecological sarcoma survey

**15. Prior to surgery, were you given any information about a possible risk of sarcoma from surgery?**
- [ ] Yes, that it may be sarcoma
- [ ] Yes, that it may be cancer
- [ ] No mention of cancer or sarcoma

**16. Did you receive support from a clinical nurse specialist (CNS)?**
- [ ] Yes - a sarcoma CNS
- [ ] Yes - a gynae cancer CNS
- [ ] No support was received

**17. Nature of surgery actually undertaken?**
- [ ] Open surgery: Total Abdominal Hysterectomy (ovaries not removed)
- [ ] Open surgery: Total Abdominal Hysterectomy/Bilateral Salpingo-Oophorectomy (ovaries removed)
- [ ] Vaginal hysterectomy
- [ ] Myomectomy (surgery to remove fibroid but preserve uterus)
- [ ] Laparoscopic procedure
- [ ] No surgery – other treatment option

**18. At what stage were you given a diagnosis of sarcoma?**
- [ ] Before surgery following imaging
- [ ] Before surgery after a biopsy
- [ ] After surgery
- [ ] Other (please specify)

**19. If you were diagnosed after surgery, how long did it take to receive a diagnosis post operation?**

**20. Was your diagnosis subsequently modified or were you given a new diagnosis? Please give details.**

**21. Had the sarcoma already spread to another part of your body at diagnosis (metastatic disease) e.g to the lungs?**
- [ ] Yes
- [ ] No

**22. Were you referred to a sarcoma specialist at this time?**
- [ ] Yes
- [ ] No

**23. If no, did your doctor mention that a sarcoma specialist was consulted to agree on a care pathway for you?**
- [ ] Yes
- [ ] No

**24. Were there additional treatment options offered to you following your initial surgery?**
- [ ] Yes
- [ ] No
  - If yes, please give details of the treatments offered

**25. If yes, what reasons were given?**
- [ ] There was some residual tumour left inside you
- [ ] Some tumour cells may have been left behind
- [ ] Some tumour cells may have spread into the pelvis
- [ ] It is possible lymph nodes may have been affected
- [ ] Microscopic tumour cells may be circulating in the bloodstream
- [ ] Other (please specify)

**26. What treatments, if any, did you have following your initially surgery?**
- [ ] Yes, radiotherapy localised to the tumour site
- [ ] Yes, radiotherapy more regional in the pelvis
- [ ] Yes, chemotherapy
- [ ] No treatment
  - Please indicate below if you did not undergo the treatment option offered to you.
27. If you received chemotherapy, what drug/s were given?

28. Was this further treatment you received overseen by a sarcoma specialist?

29. How regularly do you attend follow-up clinic?

30. Who is your follow-up with?

31. Which of the following does follow-up include?

32. Please state the frequency of monitoring e.g. chest x-ray every visit

33. Has the tumour recurred since finishing the initial course of treatment?

34. If yes, where?

35. How long after the surgery was the recurrence diagnosed?

36. If not already with a sarcoma specialist, were you referred at this stage?

37. If no, did your doctor mention that a sarcoma specialist was consulted to agree on a care pathway for you?

38. What further treatment has been given?

39. If you received chemotherapy, what drug/s were given?

40. Was this treatment under the supervision of a sarcoma specialist?

41. Has the tumour been hormone tested?

42. If yes, when and who arranged it?
43. What was the result of hormone testing?

- [ ] Positive
- [ ] Negative

44. If positive was a treatment prescribed for this (e.g. an aromatase inhibitor such as Letrozole/Anastrozole)?

- [ ] Yes
- [ ] No

45. What other treatments have you received for this cancer?

46. Was any other type of treatment embarked upon at any previous stage of your life that you now consider relevant to your current situation? I.e. medication, other surgical procedures, hormonal implants/coils etc. Please feel free to elaborate.

47. Everyone’s journey is different, and may not necessarily followed the pattern of this questionnaire, so if you wish to share any further comments about your diagnosis and treatment to help us better understand your story, please do so below.

It would be useful for Sarcoma UK to be able to contact you about your experiences. Please indicate that you would be happy to discuss this further with us by filling in your contact details below.

Name: ___________________________ Phone number: ___________________________

Email: ___________________________

Thank you!

for taking the time to complete this survey.

Sarcoma UK

The bone & soft tissue cancer charity

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This report is dedicated to all women and their families who are affected by gynaecological sarcoma.