This booklet is for people who’d like more information about Paget’s disease of the breast (also known as Paget’s disease of the nipple). It describes what Paget’s disease is, the symptoms, how it is diagnosed and how it may be treated.
This information is by Breast Cancer Care.

We are the only specialist UK-wide charity that supports people affected by breast cancer. We’ve been supporting them, their family and friends and campaigning on their behalf since 1973.

Today, we continue to offer reliable information and personal support, over the phone and online, from nurses and people who’ve been there. We also offer local support across the UK.

From the moment you notice something isn’t right, through to treatment and beyond, we’re here to help you feel more in control.

For breast cancer care, support and information, call us free on 0808 800 6000 or visit breastcancercare.org.uk
We hope this booklet helps you to ask your specialist team questions and be involved as much as you want in decisions about your treatment.

We recommend that you read this booklet alongside our booklets *Treating primary breast cancer* and *Breast cancer and you: coping with diagnosis, treatment and the future*.

What is Paget’s disease of the breast?

Paget’s disease of the breast is an uncommon type of breast cancer that usually first shows as changes to the nipple. It occurs in less than 5% of all women with breast cancer. Men can also get Paget’s disease but this is very rare.

Paget’s disease of the breast is not the same as Paget’s disease of the bone.

What are the symptoms of Paget’s disease?

The most common symptom is a red, scaly rash on the nipple, which may spread to the darker area of skin around the nipple (areola). The rash can feel itchy or you may have a burning sensation. The nipple may be pulled in (inverted). There may also be some liquid (discharge) coming from the nipple.

Paget’s disease can look like other skin conditions such as eczema or psoriasis. But there are differences. For example, Paget’s disease affects the nipple from the start while eczema generally affects the areola and only rarely affects the nipple. Also, Paget’s disease usually occurs in one breast while most other skin conditions tend to affect both breasts.
Ductal carcinoma in situ (DCIS)

Most people with Paget’s disease will have an early form of breast cancer – known as ductal carcinoma in situ (DCIS) – somewhere in the breast.

DCIS is graded as either low, intermediate or high grade, based on what the cells look like under the microscope. With Paget’s disease it’s likely to be high-grade DCIS.

DCIS means that cancer cells have developed inside the milk ducts, but remain entirely in situ (in their place of origin). They have not yet developed the ability to spread outside the ducts, either into the surrounding breast tissue or to other parts of the body.

Because it’s confined to the breast ducts, if treated DCIS has a good outlook (prognosis). However, if DCIS is left untreated, the cancer cells may develop the ability to spread from the ducts into the surrounding breast tissue and become an invasive breast cancer (see below). Although the type, size and grade of the DCIS can help predict if it will become invasive, there is currently no way of knowing for certain if this will happen.

If you have DCIS you may find it useful to read our booklet Ductal carcinoma in situ (DCIS).

Invasive breast cancer

Some people with Paget’s disease will also have developed an invasive breast cancer. In many cases this will present as a breast lump. Invasive breast cancer is breast cancer that has the potential to spread from the breast to other parts of the body. Even when there is no lump, some people may still have an invasive cancer.

For more information about invasive breast cancer see our Primary breast cancer information pack.
How is Paget’s disease diagnosed?

Because Paget’s disease is rare and can look like other skin conditions, it’s not always diagnosed straight away. Once your GP has referred you to a specialist, you may have several tests, including:

- a mammogram (a breast x-ray)
- an ultrasound scan (uses high-frequency sound waves to produce an image)

Biopsy

You will usually then have a biopsy to confirm the diagnosis. A biopsy is the removal of tissue to be looked at under a microscope. The kind of biopsy you have will depend on your symptoms. For example:

- a nipple scrape removes cells from the skin of the affected nipple
- a punch biopsy removes a small circle of tissue from the skin of the breast or nipple
- a core biopsy removes a small sample of tissue from the area of concern if this can be felt within the breast

If the area of concern can only be seen on a mammogram or ultrasound, you may have an image-guided biopsy. This is where samples of breast tissue are taken using a mammogram or ultrasound to help locate the exact position of the area of concern.

These tests can be done using a local anaesthetic. The samples of tissue or cells are sent to a laboratory where they are examined under a microscope to make a diagnosis.

If you’d like more information about the tests and procedures you may be having, see our booklet Your breast clinic appointment.
How is Paget’s disease treated?

Surgery

Surgery is usually the first treatment for Paget’s disease. The type of surgery will depend on the area of the breast affected, the size of the cancer compared to the size of your breast, and whether more than one area in the breast is affected.

You may be offered breast-conserving surgery (also called wide local excision or lumpectomy). This is the removal of the cancer with a margin (border) of normal breast tissue around it. For Paget’s disease, this type of surgery also includes the removal of the nipple and areola (the darker area of skin around the nipple).

A mastectomy (removal of all the breast tissue including the nipple area) is usually recommended if:

- the breast cancer affects a large area of the breast
- it hasn’t been possible to get a clear margin of normal tissue around the breast cancer using breast-conserving surgery
- there is more than one area of cancer in the breast
- breast-conserving surgery is not expected to provide an acceptable cosmetic result because of the position or size of the cancer

You may be offered a choice between a mastectomy and breast-conserving surgery depending on the size and location of the breast cancer within the breast. Your breast surgeon will discuss this with you and you can talk through your decision with your breast care nurse.

Breast reconstruction after Paget’s disease

If you are going to have a mastectomy, you will usually be given the option of having breast reconstruction to create a new breast shape, either at the same time as your mastectomy (immediate reconstruction) or months or years later (delayed reconstruction).

It is possible to have the nipple reconstructed after surgery for Paget’s disease of the breast, whether you have breast-conserving surgery or a mastectomy.

For more information, see our [Breast reconstruction](#) booklet.
Choosing whether or not to have breast reconstruction is a very personal decision. Some women feel reconstruction is necessary to restore their confidence. Others prefer to wear an external breast form (prosthesis), and some women choose not to have reconstruction and not to wear a prosthesis.

If you’d like more information about wearing a prosthesis, see our booklet *Breast prostheses, bras and clothes after surgery.*

**Lymph node removal**
If you have invasive breast cancer underlying the Paget’s disease, your specialist team will want to check if any of the lymph nodes (glands) under the arm (axilla) contain cancer cells. This, along with other information about your breast cancer, helps them decide whether or not you will benefit from any additional treatment after surgery. To do this, your surgeon is likely to recommend an operation to remove either some of the lymph nodes (a lymph node sample or biopsy) or all of them (a lymph node clearance).

**Sentinel lymph node biopsy**
Sentinel lymph node biopsy is widely used if tests before surgery show no evidence of the lymph nodes containing cancer cells. It identifies whether the sentinel lymph node (the first lymph node that the cancer cells are most likely to spread to) is clear of cancer cells. There may be more than one sentinel lymph node. If clear, this usually means the other nodes are clear too, so no more will need to be removed. Sentinel lymph node biopsy is usually carried out at the same time as your cancer surgery but may be done before your surgery.

If the results of the sentinel lymph node biopsy show that the first node or nodes are affected, more surgery or radiotherapy to the remaining lymph nodes may be recommended.

Sentinel lymph node biopsy is not suitable if tests before your operation show that your lymph nodes contain cancer cells. In this case it is likely that your surgeon will recommend a lymph node clearance.

Usually the lymph nodes under the arm don’t need to be removed if you have DCIS. This is because the cancer cells haven’t developed the ability to spread outside the ducts into the surrounding breast tissue. However, surgery to the lymph nodes may be recommended for some people with DCIS.
What are the adjuvant (additional) treatments?

After surgery, you may need other treatments. These are called adjuvant treatments and can include:

- chemotherapy
- radiotherapy
- hormone (endocrine) therapy
- targeted (biological) therapy
- bisphosphonates

The aim of these treatments is to reduce the risk of breast cancer returning in the same breast or developing in the opposite breast, or spreading somewhere else in the body. Some of these treatments may be given before surgery. This is known as neo-adjuvant or primary treatment.

Chemotherapy

Whether chemotherapy is recommended will depend on various features of the cancer, such as its grade, size and whether the lymph nodes contain cancer cells. Chemotherapy may be used if the underlying breast cancer is invasive. It is not used to treat DCIS. If you'd like more information see our Chemotherapy for breast cancer booklet.

Radiotherapy

Radiotherapy uses high energy x-rays to destroy cancer cells. If you have breast-conserving surgery you will usually be offered radiotherapy to the breast to reduce the risk of the cancer coming back in the same breast.

Sometimes you may be offered radiotherapy to the nodes under your arm. Radiotherapy is sometimes given to the chest wall following a mastectomy, for example if some of the lymph nodes under the arm are affected.

For more information see our Radiotherapy for primary breast cancer booklet.
Hormone (endocrine) therapy
The hormone oestrogen can stimulate some breast cancers to grow. A number of hormone therapies work in different ways to block the effect of oestrogen on cancer cells.

Hormone therapy will only be prescribed if your breast cancer has receptors within the cell that bind to the hormone oestrogen, known as oestrogen receptor positive or ER+ breast cancer. When oestrogen binds to these receptors, it can stimulate the cancer to grow.

All invasive breast cancers are tested for oestrogen receptors using tissue from a biopsy or after surgery. DCIS may be tested but this is not done in all hospitals.

If your cancer is oestrogen receptor positive, your specialist will discuss with you which hormone therapy they think is most appropriate.

Tests will also be done for progesterone (another hormone) receptors. The benefits of hormone therapy are less clear for people whose breast cancer is only progesterone receptor positive (PR+ and ER-). Very few breast cancers fall into this category. However, if this is the case for you your specialist will discuss with you whether hormone therapy is appropriate.

If your cancer is found to be hormone receptor negative, then hormone therapy will not be of any benefit to you.

If you would like more information, see our Treating primary breast cancer booklet or our individual hormone drug booklets.

Targeted (biological) therapy
This is a group of drugs that block the growth and spread of cancer. Targeted therapies target and interfere with processes in the cells that cause cancer to grow.

The most widely used targeted therapy is trastuzumab. Only people whose invasive cancer has high levels of HER2 (called HER2 positive) will benefit from having trastuzumab. HER2 is a protein that makes cancer cells grow.

There are various tests to measure HER2 levels which are done on breast tissue removed during a biopsy or surgery. If your cancer is found to be HER2 negative, then trastuzumab will not be of any benefit.
For more information see our Trastuzumab booklet or visit breastcancercare.org.uk for information about other targeted therapies.

**Bisphosphonates**

Bisphosphonates are a group of drugs that can reduce the risk of invasive breast cancer spreading in postmenopausal women. They can be used regardless of whether the menopause happened naturally or because of breast cancer treatment.

Bisphosphonates can also slow down or prevent bone damage. They’re often given to people who have, or are at risk of, osteoporosis (when bones lose their strength and become more likely to break).

Bisphosphonates can be given as a tablet (orally) or into a vein (intravenously).

Your specialist team can tell you if bisphosphonate treatment would be suitable for you.
Further support

Being diagnosed with breast cancer can make you feel lonely or isolated. There are people who can support you so don’t be afraid to ask for help if you need it.

Some people find it helpful to discuss their feelings and concerns with their breast care nurse or specialist. If you’d like to talk through your feelings and concerns in more depth over a period of time, you may want to see a counsellor or psychologist. Your breast care nurse, specialist or GP can arrange this.

Many people find it helps to talk to someone who has been through the same experience as them. Breast Cancer Care’s Someone Like Me service can put you in touch with someone who has had a diagnosis of breast cancer, so you can talk through your worries and share experiences over the phone or by email. You can also visit our confidential online Forum and join one of the ongoing discussions.

If you would like any further information and support about breast cancer or just want to talk things through, you can speak to one of our experts by calling our free Helpline on 0808 800 6000.
Four ways to get support

We hope this information was helpful, but if you have questions, want to talk to someone or read more about breast cancer, here’s how you can.

Speak to our nurses or trained experts. Call our free Helpline on 0808 800 6000 (Monday to Friday 9am–4pm and Saturday 9am–1pm). The Helpline can also put you in touch with someone who knows what it’s like to have breast cancer.

Chat to other women who understand what you’re going through in our friendly community, for support day and night. Look around, share, ask a question or support others at forum.breastcancercare.org.uk

Find trusted information you might need to understand your situation and take control of your diagnosis or order information booklets at breastcancercare.org.uk

See what support we have in your local area. We’ll give you the chance to find out more about treatments and side effects as well as meet other people like you.
Visit breastcancercare.org.uk/in-your-area
We’re here for you: help us to be there for other people too

If you found this booklet helpful, please use this form to send us a donation. Our information resources and other services are only free because of support from people such as you.

Donate today and together we can ensure that everyone affected by breast cancer has someone to turn to.

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Please accept my donation of £10/£20/my own choice of £

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In addition, we’d love to keep you updated about our work and provide you with other opportunities to get involved with Breast Cancer Care. Please tell us how you would like to hear from us (by ticking these boxes you confirm you are 18 or over)

☐ I’d like to hear from you by email
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We never give your information to other organisations to use for their own purposes. To change your preferences, or find out more information on how we use your data, please view our privacy policy at breastcancercare.org.uk or contact supporter services on 0345 092 0800.

Please return this form to Breast Cancer Care, Freepost RRKZ-ARZY-YCKG, Chester House, 1–3 Brixton Road, London SW9 6DE

Code: LP
About this booklet

Paget’s disease of the breast was written by Breast Cancer Care’s clinical specialists, and reviewed by healthcare professionals and people affected by breast cancer.

For a full list of the sources we used to research it:

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Email publications@breastcancercare.org.uk

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When you have breast cancer, everything changes. At Breast Cancer Care, we understand the emotions, challenges and decisions you face every day, and we know that everyone’s experience is different.

For breast cancer care, support and information, call us free on 0808 800 6000 or visit breastcancercare.org.uk