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 a diagnosis is made and how it is treated.
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 with our *Treating breast cancer* booklet.

**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?**

The most common symptom is a red, scaly rash involving the nipple, which may spread to the coloured 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 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.

**DCIS (ductal carcinoma in situ)**

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. This 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 within the breast or elsewhere in the body. Because it’s confined to the breast ducts, DCIS has a good outlook (prognosis).

However, if DCIS is not treated, the cancer cells may develop the ability to spread from the ducts into the surrounding breast tissue and become an invasive breast cancer. The type, size and grade of the DCIS can help
predict if it will become invasive, but currently there is no way of knowing for certain in each individual case.

**Invasive breast cancer**

Some people with Paget’s disease will also have an underlying breast lump. In most cases this will be invasive breast cancer. 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.

**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 (local doctor) has referred you to a specialist, you may have several tests to help make the diagnosis including:

- a mammogram (a breast x-ray)
- an ultrasound scan (uses high-frequency sound waves to produce an image)
- a breast MRI (magnetic resonance imaging) scan (uses magnetic fields and radio waves to produce a series of images of the inside of the breast)
- a biopsy (removal of tissue to be looked at under a microscope).

**Biopsy**

You will usually have a biopsy to confirm the diagnosis. The kind of biopsy you have will depend on your symptoms. For example:

- a punch biopsy removes a small circle of tissue from the breast or nipple
- a nipple scrape removes cells from the skin of the affected nipple
- a core biopsy removes a small sample of tissue from the area of concern that 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 can accurately locate the affected area so that a sample of tissue can be removed.
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.

What are the treatments?

Surgery
Surgery is usually the first treatment for Paget’s disease. The type of surgery will depend on the extent of the cancer within the breast.

Breast-conserving surgery, also called wide local excision or lumpectomy, 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.

A mastectomy (removal of all the breast tissue including the nipple area) is usually recommended in any of the following situations.

- 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 extent 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.

If you’re given a choice, it means that your specialist believes that you’ll get equal benefit from either treatment, and you can choose the one you feel is best for you.

If you are going to have a mastectomy, you will usually be offered breast reconstruction. This can be done at the same time as your mastectomy (immediate reconstruction) or months or years later (delayed reconstruction). It is also possible to have the nipple reconstructed. If you’d like more information, see our Breast reconstruction booklet.
Some women choose not to or cannot have a breast reconstruction. They may use a breast prosthesis (an artificial breast form) to give them the shape of a breast or may prefer not to use anything. If you’d like more information 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 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 (sentinel lymph node biopsy or lymph node sample) or all of the lymph nodes (lymph node clearance).

Sentinel lymph node biopsy is widely used for people with breast cancer whose tests before surgery show no evidence of the lymph nodes containing cancer cells. It identifies whether or not the first, or sentinel, lymph node (or nodes) is clear of cancer cells. If it is, this usually means the other nodes are clear too, so no more will need to be removed.

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

Sentinel lymph node biopsy is not suitable if tests before your operation show that your lymph nodes contain cancer cells. In this case your surgeon may 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.

For more information, see our *Treating breast cancer* and *Ductal carcinoma in situ (DCIS)* booklets.
What are the (additional) adjuvant treatments?

After surgery, you may need further treatment. This is called adjuvant (additional) therapy and can include:

- chemotherapy
- radiotherapy
- hormone therapy
- targeted therapy.

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.

Sometimes chemotherapy or hormone therapy may be given before surgery. This is known as neo-adjuvant or primary therapy.

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 carefully measured and controlled 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 may be given to the chest wall following a mastectomy in some circumstances, for example if some of the lymph nodes under the arm are affected.

It’s usually given daily (Monday to Friday) for three weeks, or every other day for five weeks, with a rest at the weekend.

Your specialist will explain the likely benefits of radiotherapy for you and also tell you about any possible side effects. For more information see our Radiotherapy for primary (early) breast cancer booklet.
**Hormone (endocrine) therapy**
As the hormone oestrogen can play a part in stimulating some breast cancers to grow, hormone therapies work 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 and stimulate the cancer to grow (known as oestrogen receptor positive or ER+ breast cancer).

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.

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 breast cancer booklet or our individual hormone drug booklets.

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

The most widely used targeted therapy is trastuzumab (Herceptin). 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 help you. For more information see our Trastuzumab (Herceptin) booklet.
Helping you face breast cancer

If you’ve been diagnosed with breast cancer there’s a lot to take in. It can be an emotional time for you, your family and friends. Our free information and support services are here to help – on the phone, or online 24 hours a day.

Ask us
Calls to our free Helpline are answered by specialist nurses and trained staff with personal experience of breast cancer. They’ll understand the issues you’re facing and can answer your questions. Or you can Ask the Nurse by email instead via our website.

Free Helpline 0808 800 6000 (Text Relay 18001)
Monday–Friday 9am–5pm, Saturday 10am–2pm
www.breastcancercare.org.uk/ATN

Expert information
Written and reviewed by healthcare professionals and people affected by breast cancer, our free booklets and other information resources cover all aspects of living with breast cancer. Download or order booklets from our website or call the Helpline.

Talk to someone who understands
Our Someone Like Me service puts you in contact with someone else who’s had breast cancer and who’s been fully trained to help. This can be over the phone or by email.

You can also chat to other people going through breast cancer on our online discussion Forum. It’s easy to use, professionally moderated and available to read any time of day.

Find out more about all of our services for people with breast cancer at www.breastcancercare.org.uk/services or phone the Helpline.
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.

We want to be there for every person facing the emotional and physical trauma of a breast cancer diagnosis. Donate today and together we can ensure that everyone affected by breast cancer has someone to turn to.

Donate by post
Please accept my donation of £10/£20/my own choice of £

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We might occasionally want to send you more information about our services and activities

☐ Please tick if you’re happy to receive email from us
☐ Please tick if you don’t want to receive post from us

We won’t pass on your details to any other organisation or third parties.

Please return this form to Breast Cancer Care, Freepost RRKZ-ARZY-YCKG, 5–13 Great Suffolk Street, London SE1 0NS
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:

Phone 0345 092 0808
Email publications@breastcancercare.org.uk

You can order or download more copies from www.breastcancercare.org.uk/publications

For a large print, Braille, DAISY format or audio CD version:

Phone 0345 092 0808
Email publications@breastcancercare.org.uk
Breast Cancer Care is the only UK-wide charity providing specialist support and tailored information for anyone affected by breast cancer.

Our clinical expertise and emotional support network help thousands of people find a way to live with, through and beyond breast cancer.

Visit [www.breastcancercare.org.uk](http://www.breastcancercare.org.uk) or call us free on 0808 800 6000 (Text Relay 18001).

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