

Paget Disease of the Breast

Key Points

- Paget disease of the breast is a rare type of cancer involving the skin of the nipple and, usually, the darker circle of skin around it, known as the areola.
- Most of the time, people with Paget disease of the breast also have one or more tumors inside the same breast.
- Paget disease of the breast may be misdiagnosed at first because its early symptoms are similar to those caused by some benign skin conditions.
- The outlook for people diagnosed with Paget disease of the breast depends on a variety of factors, including the presence or absence of invasive cancer in the affected breast and, if invasive cancer is present, whether or not it has spread to nearby lymph nodes.

1. What is Paget disease of the breast?

Paget disease of the breast (also known as Paget disease of the nipple and mammary Paget disease) is a rare type of cancer involving the skin of the nipple and, usually, the darker circle of skin around it, which is called the areola. Most people with Paget disease of the breast also have one or more tumors inside the same breast. These breast tumors are either ductal carcinoma in situ or invasive breast cancer (1–3).

Paget disease of the breast is named after the 19th century British doctor Sir James Paget, who, in 1874, noted a relationship between changes in the nipple and breast cancer. (Several other diseases are named after Sir James Paget, including Paget disease of bone and extramammary Paget disease, which includes Paget disease of the vulva and Paget disease of the penis. These other diseases are not related to Paget disease of the breast. This fact sheet discusses only Paget disease of the breast.)

Malignant cells known as Paget cells are a telltale sign of Paget disease of the breast. These cells are found in the epidermis (surface layer) of the skin of the nipple and the areola. Paget cells often have a large, round appearance under a microscope; they may be found as single cells or as small groups of cells within the epidermis.

2. Who gets Paget disease of the breast?

Paget disease of the breast occurs in both women and men, but most cases occur in women. Approximately 1 to 4 percent of all cases of breast cancer also involve Paget disease of the breast. The average age at diagnosis is 57 years, but the disease has been found in adolescents and in people in their late 80s (2, 3).

3. What causes Paget disease of the breast?

Doctors do not fully understand what causes Paget disease of the breast. The most widely accepted theory is that cancer cells from a tumor inside the breast travel through the milk ducts to the nipple and areola. This would explain why Paget disease of the breast and tumors inside the same breast are almost always found together (1, 3).



X 6 0 3 9

A second theory is that cells in the nipple or areola become cancerous on their own (1, 3). This would explain why a few people develop Paget disease of the breast without having a tumor inside the same breast. Moreover, it may be possible for Paget disease of the breast and tumors inside the same breast to develop independently (1).

4. What are the symptoms of Paget disease of the breast?

The symptoms of Paget disease of the breast are often mistaken for those of some benign skin conditions, such as dermatitis or eczema (1–3). These symptoms may include the following:

- Itching, tingling, or redness in the nipple and/or areola
- Flaking, crusty, or thickened skin on or around the nipple
- A flattened nipple
- Discharge from the nipple that may be yellowish or bloody

Because the early symptoms of Paget disease of the breast may suggest a benign skin condition, and because the disease is rare, it may be misdiagnosed at first. People with Paget disease of the breast have often had symptoms for several months before being correctly diagnosed.

5. How is Paget disease of the breast diagnosed?

A nipple biopsy allows doctors to correctly diagnose Paget disease of the breast. There are several types of nipple biopsy, including the procedures described below.

- Surface biopsy: A glass slide or other tool is used to gently scrape cells from the surface of the skin.
- Shave biopsy: A razor-like tool is used to remove the top layer of skin.
- Punch biopsy: A circular cutting tool, called a punch, is used to remove a disk-shaped piece of tissue.
- Wedge biopsy: A scalpel is used to remove a small wedge of tissue.

In some cases, doctors may remove the entire nipple (1). A pathologist then examines the cells or tissue under a microscope to look for Paget cells.

Most people who have Paget disease of the breast also have one or more tumors inside the same breast. In addition to ordering a nipple biopsy, the doctor should perform a clinical breast exam to check for lumps or other breast changes. As many as 50 percent of people who have Paget disease of the breast have a breast lump that can be felt in a clinical breast exam. The doctor may order additional diagnostic tests, such as a diagnostic mammogram, an ultrasound exam, or a magnetic resonance imaging scan to look for possible tumors (1, 2).

6. How is Paget disease of the breast treated?

For many years, mastectomy, with or without the removal of lymph nodes under the arm on the same side of chest (known as axillary lymph node dissection), was regarded as the standard surgery for Paget disease of the breast (3, 4). This type of surgery was done because patients with Paget disease of the breast were almost always found to have one or more tumors inside the same breast. Even if only one tumor was present, that tumor could be located several centimeters away from the nipple and areola and would not be removed by surgery on the nipple and areola alone (1, 3, 4).

Studies have shown, however, that breast-conserving surgery that includes removal of the nipple and areola, followed by whole-breast radiation therapy, is a safe option for people with Paget disease of the breast who do not have a palpable lump in their breast and whose mammograms do not reveal a tumor (3–5).

People with Paget disease of the breast who have a breast tumor and are having a mastectomy should be offered sentinel lymph node biopsy to see whether the cancer has spread to the axillary lymph nodes. If cancer cells are found in the sentinel lymph node(s), more extensive axillary lymph node surgery may be needed (1, 6, 7). Depending on the stage and other features of the underlying breast tumor (for example, the presence or absence of lymph node involvement, estrogen and progesterone receptors in the tumor cells, and HER2 protein overexpression in the tumor cells), adjuvant therapy, consisting of chemotherapy and/or hormonal therapy, may also be recommended.

7. What is the prognosis for people with Paget disease of the breast?

The prognosis, or outlook, for people with Paget disease of the breast depends on a variety of factors, including the following:

- Whether or not a tumor is present in the affected breast
- If one or more tumors are present in the affected breast, whether those tumors are ductal carcinoma in situ or invasive breast cancer
- If invasive breast cancer is present in the affected breast, the stage of that cancer

The presence of invasive cancer in the affected breast and the spread of cancer to nearby lymph nodes are associated with reduced survival.

According to NCI's Surveillance, Epidemiology, and End Results program, the 5-year relative survival for all women in the United States who were diagnosed with Paget disease of the breast between 1988 and 2001 was 82.6 percent. This compares with a 5-year relative survival of 87.1 percent for women diagnosed with any type of breast cancer. For women with both Paget disease of the breast and invasive cancer in the same breast, the 5-year relative survival declined with increasing stage of the cancer (stage I, 95.8 percent; stage II, 77.7 percent; stage III, 46.3 percent; stage IV, 14.3 percent) (1, 3, 8, 9).

8. What research studies are under way on Paget disease of the breast?

Randomized controlled clinical trials, which are considered the "gold standard" in cancer research, are difficult to perform for Paget disease of the breast because very few people have this disease (4, 10). However, people who have Paget disease of the breast may be eligible to enroll in clinical trials to evaluate new treatments for breast cancer in general, new ways of using existing breast cancer treatments, or strategies for preventing breast cancer recurrence.

Information about current [breast cancer treatment clinical trials](#) is available by searching NCI's list of cancer clinical trials at <http://www.cancer.gov/clinicaltrials/search>. "Paget disease" can be used as a key word to limit the search results.

Alternatively, contact NCI's Cancer Information Service at 1-800-4-CANCER (1-800-422-6237) for information about clinical trials.

Selected References

1. Harris JR, Lippman ME, Morrow M, Osborne CK, editors. *Diseases of the Breast*. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2009.
2. Caliskan M, Gatti G, Sosnovskikh I, et al. Paget's disease of the breast: the experience of the European Institute of Oncology and review of the literature. *Breast Cancer Research and Treatment* 2008;112(3):513–521. [[PubMed Abstract](#)]
3. Kanitakis J. Mammary and extramammary Paget's disease. *Journal of the European Academy of Dermatology and Venereology* 2007;21(5):581–590. [[PubMed Abstract](#)]
4. Kawase K, Dimaio DJ, Tucker SL, et al. Paget's disease of the breast: there is a role for breast-conserving therapy. *Annals of Surgical Oncology* 2005;12(5):391–397. [[PubMed Abstract](#)]
5. Marshall JK, Griffith KA, Haffty BG, et al. Conservative management of Paget disease of the breast with radiotherapy: 10- and 15-year results. *Cancer* 2003;97(9):2142–2149. [[PubMed Abstract](#)]
6. Sukumvanich P, Bentrem DJ, Cody HS, et al. The role of sentinel lymph node biopsy in Paget's disease of the breast. *Annals of Surgical Oncology* 2007;14(3):1020–1023. [[PubMed Abstract](#)]
7. Laronga C, Hasson D, Hoover S, et al. Paget's disease in the era of sentinel lymph node biopsy. *American Journal of Surgery* 2006;192(4):481–483. [[PubMed Abstract](#)]

8. Ries LAG, Eisner MP. Cancer of the Female Breast. In: Ries LAG, Young JL, Keel GE, et al., editors. *SEER Survival Monograph: Cancer Survival Among Adults: U.S. SEER Program, 1988–2001, Patient and Tumor Characteristics*. Bethesda, MD: National Cancer Institute, SEER Program, 2007. Retrieved April 10, 2012, from http://seer.cancer.gov/publications/survival/surv_breast.pdf.
9. Chen CY, Sun LM, Anderson BO. Paget disease of the breast: changing patterns of incidence, clinical presentation, and treatment in the U.S. *Cancer* 2006;107(7):1448–1458. [[PubMed Abstract](#)]
10. Joseph KA, Ditkoff BA, Estabrook A, et al. Therapeutic options for Paget's disease: a single institution long-term follow-up study. *Breast Journal* 2007;13(1):110–111. [[PubMed Abstract](#)]

Related Resources

- *Adjuvant and Neoadjuvant Therapy for Breast Cancer*
(<http://www.cancer.gov/cancertopics/factsheet/Therapy/adjuvant-breast>)
- *Cancer Clinical Trials*
(<http://www.cancer.gov/cancertopics/factsheet/Information/clinical-trials>)
- *Radiation Therapy for Cancer*
(<http://www.cancer.gov/cancertopics/factsheet/Therapy/radiation>)
- *Understanding Breast Changes: A Health Guide for Women*
(<http://www.cancer.gov/cancertopics/screening/understanding-breast-changes>)
- *Understanding Cancer Prognosis*
(<http://www.cancer.gov/cancertopics/factsheet/Support/prognosis-stats>)
- *What You Need To Know About™ Breast Cancer*
(<http://www.cancer.gov/cancertopics/wyntk/breast>)

This text may be reproduced or reused freely. Please credit the National Cancer Institute as the source. Any graphics may be owned by the artist or publisher who created them, and permission may be needed for their reuse.