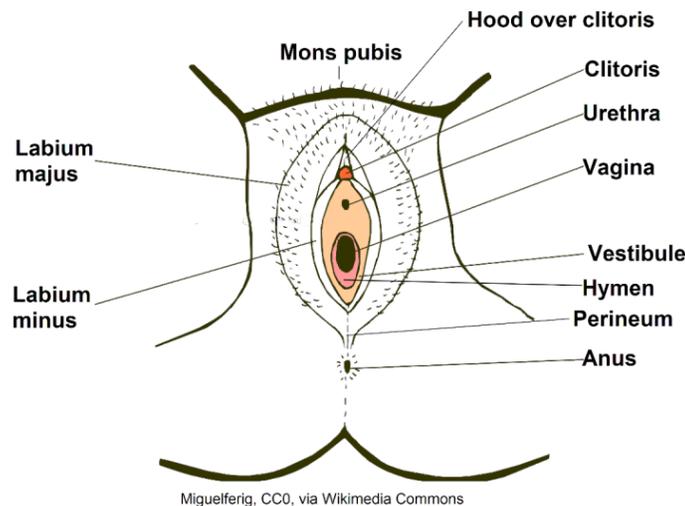


Paget's Disease of the Vulva

What is Paget's disease of the vulva?

Paget's disease of the vulva (also called **extramammary Paget's disease**) is a rare skin cancer that affects the top layers of skin. It may spread to the anus and inner thighs. While the majority of vulvar Paget's disease is precancerous, 10% of patients with vulvar Paget's disease have cancer. Up to 25% of patients with anal Paget's disease have cancer.

- The **vulva** is the part of the body that includes all the external (outside the body) genital parts. It covers the area from where pubic hair grows (the **mons pubis**) to the opening that stool comes out of (the **anus**). The vulva includes the large outer lips of skin (**labia majora**), small inner lips of skin (**labia minora**), the clitoris, the opening that urine comes out of (**urethra**), and the opening to the vagina.



What are the symptoms of Paget's disease of the vulva?

The most common symptom is an itchy rash with areas of thickened, red-colored skin that may look velvety. The skin may have moist areas that bleed easily or look scaly or crusty.

How is Paget's disease of the vulva diagnosed?

Because it can be hard to make a diagnosis based only on how the skin looks, we will remove a small piece of skin for lab testing (this is called a **biopsy**). We will give you an injection (shot) of numbing medicine before the skin is removed.

About 1 out of 4 people with this condition will also have another type of cancer. We will also test you for the possibility of another cancer type. This may include:

- Colonoscopy (using a tiny, flexible camera to look at your rectum and large bowel)
- Cystoscopy (using a tiny, flexible camera to look at your bladder)
- Mammogram
- Pap test
- Ultrasound of the pelvis
- Urine tests

How is Paget's disease of the vulva treated?

Surgery is the standard treatment when you are first diagnosed with this condition. There are different types of surgery that we can use to remove the disease. These include:

- **Wide local excision:** Removing the abnormal tissue and some normal tissue around it
- **Vulvectomy:** Removing part or all of the vulva

- Skin repair with a flap from nearby skin, or a skin graft taken from another part of your body

When deciding on the type of surgery, we will consider the size and location of your disease and talk with you about the plan. However, because this skin disease usually extends into what looks like normal skin, it can be hard to know how much tissue we need to remove. Surgery often requires a hospital stay and several weeks of recovery at home. We may ask surgeons from the Department of Plastic Surgery or our Gynecologic Oncology Division to join us in doing your surgery.

What kind of follow-up do I need to do after treatment?

- It is common for the disease to come back at or near the surgical site. We will see you for follow-up exams, typically every 4-6 months for 10 years after your surgery. During these check-ups, we will use a **vulvoscope** (using magnification to look closely at your vulva).
- Patients with Paget’s disease of the vulva have a higher chance of cancer in other parts of the body, so it is important to get routine cancer screening like mammograms and Pap tests.

Where can I find more information on Paget’s disease of the vulva?

Resource	QR code
Extramammary Paget’s disease (EMPD) Support Group www.myempd.com/support	
International Society for the Study of Vulvovaginal Disease www.issvd.org/resources/extra-mammary-pagets-disease	

Resource	QR code
<p>DermNet™ NZ was started in 1996 by a group of dermatologists from New Zealand. It has become a world-wide resource of skin disease information.</p> <p>dermnetnz.org/topics/extramammary-paget-disease</p>	

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