

Patient Information Leaflet Paget's Disease of the Vulva

What is Vulval Paget's Disease?

Paget's disease of the vulva is rare.

It is caused by an abnormal change in the cells which cover the skin (outer layer) of the vulva. It usually only affects women who've been through the menopause.

What causes Vulval Paget's Disease?

The cause of Paget's disease remains unknown.

What are the symptoms of Vulval Paget's Disease?

Symptoms may have been present for several years before seeking medical advice. The most common symptom is a mild to intense itching of a lesion found around the groin, genitalia, perineum or anal area.

Pain and bleeding may occur from scratching lesions that have been around for a long time.

Thickened plaques may form that can become red, scaly and crusty. Although they may appear similar to eczema, they fail to clear up with topical steroid creams.

Can there be any complications from Vulval Paget's Disease?

Vulval Paget's is most often an in-situ tumour of the apocrine glands. However, vulval Paget disease may be associated with an underlying cancer, either in the vulval area, or a more widespread invasive cancer.

How is Vulval Paget's Disease diagnosed?

The diagnosis is made by removing a small piece of tissue (biopsy) which is then examined under a microscope. The biopsy is done to confirm the diagnosis and exclude other conditions. Investigations to search for nearby cancers may also be done.

What is the treatment for Vulval Paget's Disease?

The mainstay treatment of vulva Paget's disease is surgery to remove the area and a wide margin of clear tissue (wide local excision). Often however several treatments may be used.

Photodynamic therapy (the combination of a cream to make the Paget's cells sensitive to light and a specific wave length of light) is currently being studied. Imiquimod (a cream applied to the skin and used for another pre-invasive vulval condition) is also being prescribed as a treatment.

Radiotherapy may also be used to treat vulval Paget's disease.

How often will I need to attend clinic?

It's not unusual for Paget's disease of the vulva to come back again. Because of this women are carefully followed up and seen regularly by their gynaecologists for the long term. Women should be re-examined every 3 months for 2 years after initial diagnosis, after which annual follow-ups are recommended.

Where can I get further information about Vulval Paget's Disease?

We recommend that you use dedicated websites or helplines to gain further information on lichen sclerosus as generic internet searches can lead to you reading information that is not accurate or out of date.

Vulval Paget's Support Group

- <http://www.vulvapagetssupport.org>.