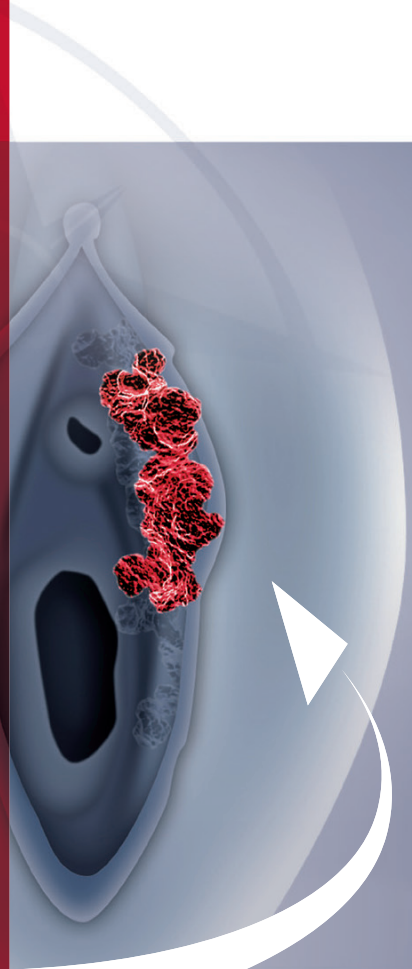


Vulvar Cancer Guidelines

SUMMARY
FOR PATIENTS
AND PUBLIC



VULVAR CANCER GUIDELINES

SUMMARY FOR PATIENTS AND PUBLIC

*The European Society of Gynaecological Oncology (ESGO)
is a non-profit organization dedicated to management
and prevention of gynaecological cancer.
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Your support will be appreciated and acknowledged.*

Dear patient,

This document sets out the adapted guidelines for the treatment of vulvar cancer. This is an up-to-date adaption for lay people (patients, but also a patient's relatives and friends) that has been modified from the European Society for Gynaecological Oncology (ESGO) professional guidelines that your medical team use when managing vulvar cancer.

It may be that, despite the adaptation, the technical content and terms will be difficult to understand fully. However, our aim is to show you what a professional guideline is like. Excessive simplification, which would make things look "simple", has been avoided.

We hope that after reading this you will understand why your doctors ask you certain questions, administer various tests, discuss results, change decisions, and advise you to undergo or to refuse particular medical procedures. You will also see how strong and specific these recommendations sometimes are. You will see how tremendous knowledge and experience need to come together so that your medical team can foresee all the factors, including complicated ethical and psycho-social factors, needed to avoid clinical or technical errors.

On the other hand, recommendations are not always adapted to individual cases. In addition, guidelines from other international or national learned societies are available. Whatever the guidelines, your doctor is not obliged to follow them, but must show knowledge of them, and must explain to you the reasoning if he or she does not strictly follow them. You will see how difficult it can be to maintain an individualised, personalised attitude while simultaneously following the approved recommendations.

Finally, modern medicine is a close collaboration between doctor and patient. We must make common decisions to fight for your health, taking into account each other's opinions, experiences, feelings, and conditions. So, this adapted version of the professional guidelines is not only simple educational material about the disease but a real tool that can improve your understanding of the very serious work you and your doctor's team are undergoing to save your life and hopefully defeat the cancer definitively.

Sincerely,

*Eka Sanikidze (GE), Dina Kurdiani (GE), Murat Gultekin (TR),
Karina Dahl Steffensen (DK), Esra Urkmez (USA), Denis Querleu (FR),
Kim Hulscher (NL), Linda Snoep (NL)*

The European Society of Gynaecological Oncology (ESGO) developed guidelines covering diagnosis and referral, preoperative investigations, surgical management, procedures for sentinel lymph node (the first lymph node or group of nodes that show signs of cancer), radiation therapy, chemoradiation, systemic treatment, treatment of recurrent disease, and follow-up for patients with vulvar cancer.

The objective of these guidelines is to improve and to standardise the management of patients with vulvar cancer. The guidelines are intended for use by gynaecological oncologists, general gynaecologists, surgeons, pathologists, radiotherapists, medical and clinical oncologists, general practitioners, palliative care teams, and allied health professionals and are adapted here for patients to understand professionals' decisions better.

These guidelines apply to adults over the age of 18 who have squamous cell carcinoma (a type of skin cancer that begins in the squamous cells—outermost layer—of the skin) of the vulva. These guidelines do not address patients with other vulvar cancer histologies. Any clinician seeking to apply or consult these guidelines is expected to use independent medical judgment in the context of individual clinical circumstances to determine any patient's care or treatment.

The European Society of Gynaecological Oncology (ESGO www.esgo.org) is a non-profit organization dedicated to the treatment and prevention of gynaecologic cancers. If you wish to donate, please contact adminoffice@esgo.org. Your support will be appreciated and acknowledged.

The European Network of Gynaecological Cancer Advocacy Groups (ENGAGe) is a network of European patient advocacy groups established by ESGO in 2012, representing all gynaecological cancers, particularly ovarian, endometrial, cervical, vulvar, and rare cancers.

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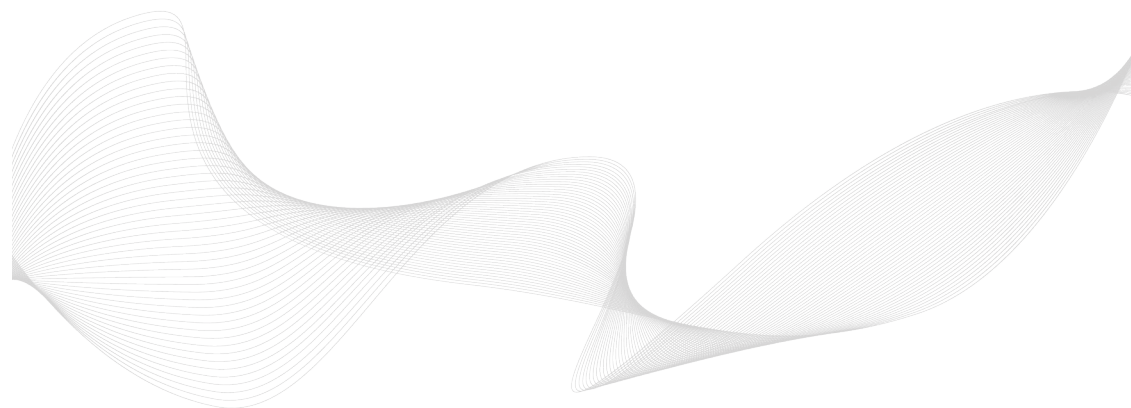
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1. PREOPERATIVE INVESTIGATIONS

For any patient for whom vulvar cancer is suspected, diagnosis should be established by a punch/incision biopsy (in which only a portion of the tumour is removed). Excision biopsy (when the entire tumour is removed) should be avoided for initial diagnosis, as this may obstruct further treatment planning.

- ✓ In patients with multiple vulvar lesions, all lesions should be biopsied separately (with clear documentation of mapping).
- ✓ All patients with vulvar cancer should be referred to a Gynaecological Oncology Centre (GOC) and treated by a multidisciplinary gynaecological oncology team.

Because vulvar cancer is a serious oncological disease, it should be diagnosed and treated properly according to the stage of cancer, the area in which the cancer has spread, and the type of lesions present. Vulvar cancer patients should not be referred to an ordinary gynaecological department, but to a Gynaecological Oncology department or centre where they can be treated by a multidisciplinary gynaecological oncology team, including a gynaecological oncologist, urologist, abdominal oncologist, clinical oncologist, chemo-/radiotherapy specialists, etc., depending on the organs involved and any concomitant diseases.

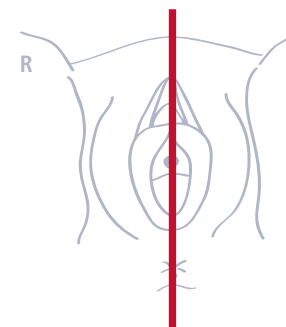


2. STAGING SYSTEM

When doctors diagnose a patient with cancer, they use a scoring system to assess the spread of the cancer, if any, and assign it to a stage. This stage is then referenced to help the medical care team plan treatment. One commonly used staging guide for vulvar cancer is the scoring system presented by the International Federation of Gynecology and Obstetrics (abbreviated **FIGO**). See www.figo.org for more information. Another system is the **TNM** Classification from the Union for International Cancer Control (UICC). See www.uicc.org for more information.

3. PREOPERATIVE INVESTIGATIONS

✓ The preoperative work-up should at least include clear documentation of the clinical exam (size of lesion, distance to the midline/clitoris/anus/vagina/urethra, and palpation of lymph nodes). A photograph or clinical drawing is advised (see below).



✓ Evaluation of the cervix/vagina/anus is recommended.

Prior to sentinel lymph node biopsy, clinical examination and imaging of the groin (either by ultrasound, positron emission tomography (PET), computed tomography (CT), or magnetic resonance imaging (MRI)) are required to identify potential lymph node metastases.

✓ Suspicious nodes (at palpation and/or imaging) should be analysed by FNA-fine-needle aspiration (when a thin needle is inserted into an area of abnormal-appearing tissue or body fluid to collect a sample for diagnosis), or core biopsy (a procedure where a needle is passed through the skin to take a sample of tissue from a mass or lump) when this would alter primary treatment.

✓ Further staging with CT thorax/abdomen and pelvis is recommended where there is proven or at least clinical suspicion of (nodal) metastatic disease and/or advanced-stage disease.

✓ The pathology report on preoperative biopsy should at least include histological type (the type of tissue in which the cancer originates) and depth of invasion.

4. SURGICAL MANAGEMENT

Local treatment

Radical local excision (surgical removal) is recommended.

- ✓ The doctor should consider an additional, more superficial resection of differentiated vulvar intraepithelial neoplasia (a condition in which abnormal cells are found on the surface of or in the tissue that lines an organ) in addition to radical local excision of invasive tumours.
- ✓ In multifocal (having more than one focus) invasive disease, radical excision of each lesion as a separate entity may be considered. Vulvectomy (partial or complete removal of vulva) may be required in cases with multifocal invasion arising on a background of extensive vulvar dermatosis.
- ✓ The goal of excision is to obtain tumour-free pathological margins. Surgical excision margins of at least 1 cm are advised. It is acceptable to consider narrower margins where the tumour lies close to midline structures (clitoris, urethra, anus), and preservation of their function is desired.
- ✓ When invasive disease extends to the pathological excision margins of the primary tumour, re-excision is the treatment of choice.
- ✓ The optimal management of the groin (full inguinofemoral lymphadenectomy—the surgical removal of superficial inguinal lymph nodes and deep femoral lymph nodes or isolated removal only) for enlarged, proven metastatic nodes remains to be defined.

Groin treatment

- ✓ Advanced-stage patients should be evaluated in a multidisciplinary setting to determine the optimal choice and order of treatment modalities.
- ✓ Where enlarged (> 2 cm) pelvic nodes are identified, their removal should be considered.

Reconstructive surgery

Availability of reconstructive surgical skills as part of the multidisciplinary team is required in early as well as advanced-stage disease.

5. SENTINEL LYMPH NODE PROCEDURE

The sentinel lymph node procedure is recommended in patients with unifocal cancers of < 4 cm, without suspicious groin nodes.

Use of radioactive tracer is mandatory; use of blue dye is optional.

Lymphoscintigram (the technical name for the various procedures used to study the lymphatic system) is advised to enable the preoperative identification, location, and number of sentinel lymph nodes.

6. RADIATION THERAPY

Adjuvant radiotherapy (radiotherapy delivered after primary treatment to destroy remaining cancer cells) should start as soon as possible, preferably within 6 weeks of surgical treatment.

- ✓ When invasive disease extends to the pathological excision margins of the primary tumour, and further surgical excision is not possible, postoperative radiotherapy should be performed.
- ✓ In case of close but clear pathological margins, postoperative vulvar radiotherapy may be considered to reduce the frequency of local recurrences. There is no consensus for the threshold of pathological margin distance below which adjuvant radiotherapy should be advised.
- ✓ Postoperative radiotherapy to the groin is recommended for cases with > 1 metastatic lymph node and/or the presence of extracapsular lymph node involvement (infiltration of cancer cells beyond the capsule of the metastatic lymph node).

7. CHEMORADIATION

Definitive chemoradiation (with radiation dose escalation) is the treatment of choice in patients with unresectable disease.

In advanced-stage disease, neoadjuvant chemoradiation (delivered before the main treatment to help reduce the size of a tumour or kill cancer cells that have spread) should be considered in order to avoid exenterative surgery (radical surgical treatment that removes all organs from a person's pelvic cavity—bladder, urethra, rectum, and anus).

Radiosensitising chemotherapy (which makes tumour cells more sensitive to radiation therapy), preferably with weekly cisplatin, is recommended.

8. SYSTEMIC TREATMENT

Data in vulvar cancer are insufficient to recommend a preferred schedule in a palliative setting.

9. TREATMENT OF RECURRENT DISEASE

Treatment of vulvar recurrence

Radical local excision is recommended

✓ For vulvar recurrence with a depth of invasion > 1 mm and previous sentinel lymph node removal only, inguino-femoral lymphadenectomy should be performed.

✓ The indications for postoperative radiotherapy are comparable to those for the treatment of primary disease.

Treatment of groin recurrence

Restaging by CT (or PET-CT) of the thorax/abdomen/pelvis is recommended.

✓ The preferred treatment is radical excision when possible, followed by postoperative radiation in radiotherapy-naïve patients.

✓ The addition of radiosensitising chemotherapy to postoperative radiotherapy should be considered.

✓ Definitive chemoradiation is preferred when surgical treatment is not possible.

Treatment of distant metastases

Systemic (palliative) therapy may be considered in individual patients (see systemic treatment).

10. FOLLOW-UP

The optimal follow-up schedule for vulvar cancer is undetermined.

1. After primary surgical treatment, a follow-up schedule is suggested:

- First follow-up, 6-8 weeks postoperative
- First two years, every 3-4 months
- Third and fourth year, biannually
- Afterward, long-term follow-up, especially in cases of predisposing vulvar disease

Follow-up after surgical treatment should include clinical examination of vulva and groin.

2. After definitive (chemo)radiation, the following follow-up schedule is suggested:

- First follow-up visit, 10-12 weeks after completion of definitive (chemo)radiation
- First two years, every 3-4 months
- Third and fourth year, biannually
- Afterward, long-term follow-up, especially in case of predisposing vulvar disease

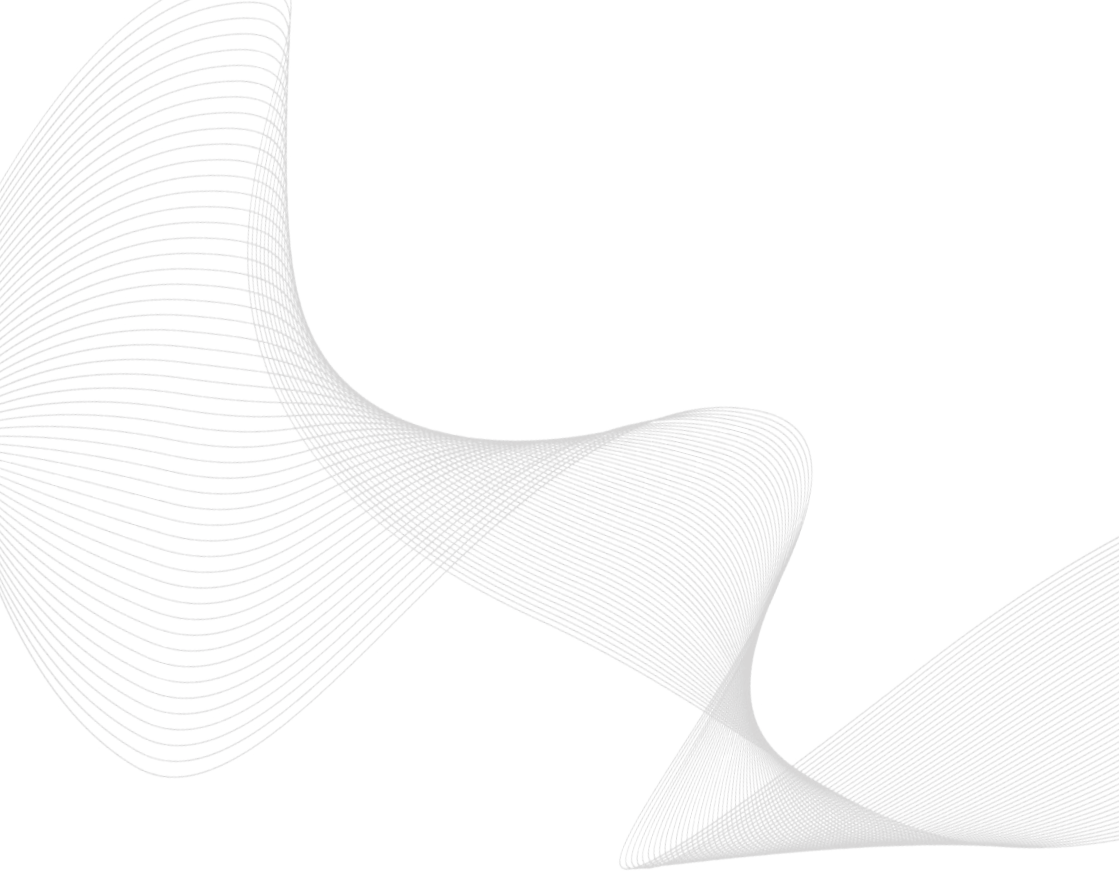
At the first follow-up visit 10-12 weeks after definitive (chemo) radiation, CT or PET-CT is recommended to document complete remission.



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and ENGAGe Executive Group members for their work
and constant availability.*

ENGAGe Contact:

<http://engage.esgo.org/>
engage@esgo.org



Access the full ESGO Guidelines



ESGO Office
7, Rue François-Versonnex
1211 Geneva 6, Switzerland
Email: adminoffice@esgomail.org
www.esgo.org