Cancer National Specialist Advisory Group

All Wales Guidelines for the Management of Vulval Cancer

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Acknowledgments

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A description of the methodology can be found in Appendix 1.

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Foreword

Vulval cancer is an uncommon condition, affecting around 80 women in Wales each year. Although the incidence is highest in the over 75 age group, it can affect women of any age. The rarity of cancer of the vulva means that there are relatively few clinical trials which may be used to direct patient care. Nevertheless, the treatment for vulval cancer is highly complex requiring specialised multidisciplinary teams to deal with the challenges of radical surgery and wound healing, chemotherapy and radiotherapy, and psychological support.

In preparing this guideline, specialists from around Wales have come together to form a consensus document which can be used by those who plan and deliver gynaecological cancer services. The aim of the guideline is to help improve and coordinate care and bring about uniformity of treatment for women with vulval cancer in Wales.

Recently the Royal College of Obstetrician and Gynaecologists has produced Guidelines for the Diagnosis and Management of Vulval Carcinoma (2014), which has been incorporated into this Welsh guideline and expanded upon to include holistic care, role of CNS, lymphoedema, and referral pathways within a Welsh context.

Louise Hanna
Chair of the Cancer NSAG Gynaecology subgroup
1. **Background**

1.1. **Incidence, age and mortality**

Vulval cancer is rare, accounting for only 3-5% of all female genital tract cancers with 54 cases in 2012 in Wales (Welsh Cancer Intelligence and Surveillance Unit data). Vulval cancer is commonest amongst women over 65 years old.

The five year survival in cases with no lymph node involvement is in excess of 80%. This falls to less than 50% if the inguinal nodes are involved and 10-15% if the iliac or other pelvic nodes are involved (Royal College of Obstetrician and Gynaecologists, 2006).

When following these guidelines the rarity of vulval cancer and lack of strong evidence should be taken into account.

1.2. **Aetiology**

Lichen sclerosus and infection with high risk types of Human papillomavirus (HPV) are both conditions in which squamous neoplastic change can be seen. There may be an intraepithelial stage seen first, i.e. vulval intraepithelial neoplasia (VIN), with either condition. Typically HPV-related disease is seen in younger women and may be multifocal. Either disease may also be present in the perianal region. Other pre-invasive conditions include Paget’s disease (adenocarcinoma in situ) and melanoma in situ.

1.3. **Pathology**

Most vulval carcinomas are of squamous cell type (SCC) (90%). SCCs include the rare verrucous carcinoma which is relatively slow growing. Three percent are malignant melanomas and the remainder is made up of basal cell carcinoma, invasive adenocarcinoma, adenoid cystic carcinoma, sarcomas and others.
Malignant melanoma should be separately reported and staged according to the system for cutaneous melanomas. Melanomas can be melanotic or amelanotic and are extremely aggressive.

Primary adenocarcinoma and adenoid cystic carcinoma most commonly arise from Bartholin’s glands. Such tumours represent 1-3% of all vulval carcinomas. They commonly involve groin and pelvic nodes with a correspondingly poor survival (52% at 5 years)(Cardosi, 2001). Sarcoma of the vulva is rare and has a tendency to local blood borne spread. Basal cell carcinoma and verrucous carcinoma are squamous variants, rarely associated with lymph node metastases (Piura, 1999).

Core minimal data for reporting: (Royal College of Pathologists, 2010):
- Tumour type, according to the WHO classification
- Tumour differentiation
- Tumour size (in at least two dimensions)
- Thickness/depth of invasion
- Presence or absence of lymphovascular invasion
- Status of all resection margins
- Minimum tumour free margins
- Presence of associated VIN or Paget’s disease
- Status of resection margins for VIN or Paget’s disease
- Minimum distance to margins for VIN or Paget's disease
- Presence or absence of non-neoplastic epithelial disease
- Presence or absence of lymph nodes metastases
- Presence of extranodal spread
- Whether nodal metastasis is larger than 5 mm

1.4. Symptoms and signs

Most women present with vulval itching, irritation or pain. Women may also notice a lump, bleeding or discharge.
2. Staging

Staging of vulval cancer is according to FIGO staging system (see table 1). Malignant melanomas are staged according to Breslow’s depth of invasion (Breslow, 1970).

<table>
<thead>
<tr>
<th>FIGO Stage (2009)</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Tumour confined to the vulva</td>
</tr>
<tr>
<td>IA</td>
<td>Lesions ≤2 cm in size, confined to the vulva or perineum and with stromal invasion ≤1.0 mm*, no nodal metastasis</td>
</tr>
<tr>
<td>IB</td>
<td>Lesions &gt;2 cm in size or with stromal invasion &gt;1.0 mm*, confined to the vulva or perineum, with negative nodes</td>
</tr>
<tr>
<td>II</td>
<td>Tumour of any size with extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with negative nodes</td>
</tr>
<tr>
<td>III</td>
<td>Tumour of any size with or without extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with positive inguino-femoral lymph nodes</td>
</tr>
<tr>
<td>IIIA</td>
<td>(i) With 1 lymph node metastasis (≥5 mm), or</td>
</tr>
<tr>
<td></td>
<td>(ii) 1–2 lymph node metastasis(es) (&lt;5 mm)</td>
</tr>
<tr>
<td>IIIB</td>
<td>(i) With 2 or more lymph node metastases (≥5 mm), or</td>
</tr>
<tr>
<td></td>
<td>(ii) 3 or more lymph node metastases (&lt;5 mm)</td>
</tr>
<tr>
<td>IIIC</td>
<td>With positive nodes with extracapsular spread</td>
</tr>
<tr>
<td>IV</td>
<td>Tumour invades other regional (upper 2/3 urethra, upper 2/3 vagina), or distant structures</td>
</tr>
<tr>
<td>IVA</td>
<td>Tumour invades any of the following:</td>
</tr>
<tr>
<td></td>
<td>(i) upper urethral and/or vaginal mucosa, bladder mucosa, rectal mucosa, or fixed to pelvic bone, or</td>
</tr>
<tr>
<td></td>
<td>(ii) fixed or ulcerated inguino-femoral lymph nodes</td>
</tr>
<tr>
<td>IVB</td>
<td>Any distant metastasis including pelvic lymph nodes</td>
</tr>
</tbody>
</table>

*The depth of invasion is defined as the measurement of the tumour from the epithelial stromal junction of the adjacent most superficial dermal papilla to the deepest point of invasion.
3. Screening

There is no screening procedure for vulval cancer. Patients with carcinoma of the vulva are at an increased risk of developing other cancers of the genital tract (Palmer, 2010), and therefore should be informed and advised to seek medical attention if new symptoms develop.

4. Referral pathways/networks

4.1. General Practitioner

If cancer of the vulva is suspected then referral should be to the nearest Cancer Unit or Centre and specifically to a gynaecologist who has additional training in oncology.

4.2. Non-oncological gynaecologists

If a patient with vulval cancer is seen by a gynaecologist in a Cancer Unit then referral should be made to the Cancer Centre as soon as the diagnosis is confirmed or strongly suspected. The rarity of the cancer, the variety of possible management techniques and the additional skills required mandate that this cancer should be managed by specialist teams. Referral should include sending all relevant histopathological material to the specialist gynaecological pathologist in the gynaecological cancer centre.

4.3. Cancer Centre

The patient should be diagnosed and treated according to the cancer waiting times targets. All new cases of vulval cancer should be discussed at the Cancer Centre multidisciplinary team meeting and the histopathological material reviewed by a specialist gynaecological pathologist, prior to radical surgery.
5. Diagnosis

Clinical features strongly indicating vulval cancer include an irregular, fungating mass, an irregular ulcer or enlarged groin nodes. If vulval cancer is strongly suspected the patient should be referred urgently to a Cancer Centre.

Any change in vulval or vaginal epithelium in a post-menopausal woman warrants a biopsy. These changes include; a swelling, polyp or lump, an ulcer, colour change (whitening or pigment deposition), elevation or irregularity of the surface contour.

Any ‘warts’ in a post-menopausal woman or persistent ‘warts’ in the pre-menopausal woman should be biopsied. In pre-menopausal women all other vulval signs and symptoms should be managed as for those in post-menopausal woman unless there is a confirmed infection. Abnormalities should be biopsied rather than excised. If cancer is confirmed, the patient should be referred to a gynaecological cancer centre. The site of biopsy/excision should be recorded on a diagram/photograph.

All cases of suspected vulval cancer should have the diagnosis confirmed with a biopsy prior to definitive treatment.

6. Pre-invasive disease

Colposcopy is useful for localisation but the signs are non-specific. Diagnosis is by biopsy. Multiple biopsies may be required to map the extent of disease and exclude invasion. VIN, Paget’s disease and melanoma-in-situ may be treated with local excision: groin node dissection is inappropriate. Women with VIN, Paget’s disease or melanoma-in-situ should be followed up by either specialist vulval clinics or gynaecological oncologists. Consideration should be given to enrolment in an appropriate trial.
7. Investigations

When considering investigations, it should be remembered that patients often present in their 7th decade and there may be appreciable medical problems. These must be corrected or adequately controlled before major surgery is contemplated. Operability rates of 96% have been reported in large centres (Monaghan and Hammond, 1984).

7.1. Pre-operative investigations for Women with vulval cancer

Investigations:
- Vulval biopsy
- Chest X-ray
- Cervical smear, if not up to date with national programme
- Full blood count, biochemical profile, liver function tests
- Abdominal and pelvic CT scan or MRI scan in stage 1b or above (for concurrent pelvic pathology and retroperitoneal nodes)
- Biopsy of any grossly involved nodes or other metastases
- Group and save/crossmatch blood sampling as appropriate
- Midstream specimen of urine (MSU)
- ECG if over 50 years

8. Treatment of primary disease

Women with vulval cancer should be treated in the Cancer Centre. Each case should be considered individually and an agreed plan of management devised by the gynaecological cancer MDT. Factors such as tumour size, location, medical fitness and the wishes of the patient will all influence management.
8.1. Treatment of early vulval cancer

A summary of this information in found in table 2.

**Surgery**

**Squamous carcinoma Stage 1A**

Small tumours with a depth of invasion of \( \leq 1 \text{mm} \) (Stage 1A) should usually be managed surgically. Surgery to the primary tumour should be radical to remove the tumour yet conservative to avoid unnecessary surgical and psychological morbidity. Wide radical local excision with a minimum margin of 1cm of disease free tissue is often sufficient (Hacker and Van der Velden, 1993). In these cases, it is not necessary to perform a groin node lymphadenectomy as the risk of involved nodes is negligible. *(Level 2)*

**Squamous carcinoma Stage 1b**

When the tumour is Stage 1B or worse a groin node assessment is also undertaken with radical local excision or as part of a radical vulvectomy (Sedlis et al., 1987). Groin node assessment may involve either excision of sentinel nodes or complete groin node dissection. Rarely primary radiotherapy to the groins may be appropriate.

**Radical vulvectomy**

Radical vulvectomy involves removal of the vulva extending from the vaginal introitus to the outer borders of the labia majora and removal of the superficial and deep inguinal (groin) nodes either with a triple incision where the groin nodes are removed separately or *en masse* with the primary tumour as a ‘butterfly’ incision. The excision should have at least a 1.5cm disease free margin around the primary tumour in the fresh specimen, to allow for tissue shrinkage (Hacker and Van der Velden, 1993; Heaps et al., 1990). *(Level 3)*
**Groin node dissection**

The triple incision approach is associated with significantly less operating time, less blood loss, shorter hospital stay and less wound morbidity (Hacker *et al.*, 1981). It also produces a less disfiguring scar. Patients with fungating groin nodes should be considered to have the groin dissection performed in continuity with the vulva, but often depends on the position of the primary tumour.

Bilateral groin node dissection is usually required because of the cross-over of lymph channels. However in very lateral tumours (the medial edge of the tumour must be at least 2cm lateral to the midline of the vulva), only an ipsilateral groin node dissection need initially be performed (Stehman *et al.*, 1992). If the ipsilateral nodes are subsequently shown to be positive for cancer, the contralateral nodes should also be excised or irradiated as the nodes are more likely to be positive.

It is recommended that the deep femoral nodes as well as the superficial groin nodes be removed. Superficial groin node dissection alone is associated with a higher risk of groin node recurrence (Stehman *et al.*, 1992). Preservation of the long saphenous vein is reported to reduce both groin wound and subsequent lower limb problems (Zhang *et al.*, 2000; Dardarian *et al.*, 2006; Zhang *et al.*, 2007). (Level 3)

Following inguinofemoral lymphadenectomy, sartorius muscle transposition may be of benefit in preventing subsequent femoral vessel damage, particularly in those women who are thin and in those for whom adjuvant groin radiation therapy is anticipated (Paley *et al.*, 1997). Use of prophylactic antibiotics and length of time of surgical drains depends on local microbiology advice and the surgeon’s discretion.

Refer to the latest NICE Guidance for thromboembolic disease prophylaxis (see [http://pathways.nice.org.uk/pathways/venous-thromboembolism#content=view-info%3Asource-guidance](http://pathways.nice.org.uk/pathways/venous-thromboembolism#content=view-info%3Asource-guidance))
Pelvic node dissection is rarely performed as 15-20% of those with involved inguinal nodes will have involved pelvic nodes and surgery may only salvage 10% of these. Radiotherapy offers better prospect of sterilising pelvic nodes with lower morbidity (Homesley et al., 1986). (Level 2)

Sentinel node excision
Evidence suggests that unifocal tumours with a maximum diameter of less than 4 cm may be safely managed by excision of sentinel lymph nodes. The 2014 RCOG guidelines recommend that this should ideally be carried out within a clinical study such as GROINS-V II and emphasise that all surgeons undertaking the procedure should do so after appropriate training and demonstration of competency. It is suggested that this should be limited to centres that have an adequate volume of work. If a sentinel node cannot be identified or contains metastatic disease, then a full groin node dissection should be carried out. All sentinel node dissections should be carried out according to GROINS-V II protocol, or similar evidence based protocol.

Primary radiotherapy to groins
Surgery should be considered as the primary treatment option for groin nodes. Morbidity after surgical treatment and groin node dissection is considerable. Primary radiotherapy to the groin is expected to result in less morbidity, however studies to date on the efficacy of primary radiotherapy to the groins in terms of groin recurrences and survival show conflicting results, surgery is still the cornerstone of therapy. Primary radiotherapy should only be carried out as part of a clinical trial, unless there are specific clinical reasons and these are documented in the case notes.

Complications of surgery
Complications have been reported to include (Wills, 2013):
- Wound disruption and infection
• Thromboembolic disease
• Secondary haemorrhage
• Leg oedema
• Femoral nerve damage
• Osteitis pubis
• Hernia
• Urinary infection
• Introital scarring and vaginal prolapse
• Changes in sexual function and body image.

Postoperative radiotherapy

Postoperative radiotherapy to the primary site

The incidence of vulval recurrence locally is related to the measured disease free surgical margin as measured in the histopathological specimen. The risk of recurrence increases as the disease free margin decreases (>8mm 0%; 8-4.8mm 8%; <4.8mm 54%) (Heaps et al., 1990). Postoperative radiotherapy to the primary site is therefore not recommended if the resection margin is greater than 8mm. In patients with close margins, further surgical excision can be considered if it is known which margin is close and if function is not going to be unduly compromised, however there is no evidence to support this.

Although a reduction in local recurrence has been shown following adjuvant local therapy in patients with close surgical margins, this was not associated with an improvement in survival. Adjuvant treatment for positive margins was shown to improve survival compared with observation alone (Faul et al., 1997). (Level 4)

Postoperative radiotherapy to the nodes

One randomised trial of patients who underwent radical vulvectomy and bilateral groin node dissection has shown a survival advantage to those given postoperative radiotherapy to the nodes compared with those given a pelvic node dissection particularly in those with two or more positive groin nodes (Homesley et al., 1986).
Postoperative radiotherapy to the loco-regional nodes should be considered when two or more lymph nodes are involved with metastatic disease or when there is complete replacement or extra capsular spread in any node. *(Level 2)*

Although both lymphovascular invasion and infiltrative growth patterns are associated with a worse prognosis, currently, adjuvant radiotherapy is not recommended in these situations, in the absence of other risk factors.

### 8.2. Treatment of advanced vulval cancer

This group includes all women with grossly involved nodes and those who have vulval lesions where there is an extensive vulval lesion necessitating functional disability for its removal.

**Surgery**

The size and location of the tumour will influence the surgical approach. Some tumours will require a radical ano-vulvectomy.

Groin node dissection should be undertaken when there are clinically suspicious groin nodes present. In cases with large primary lesions and clinically suspicious nodes, a radical vulvectomy with 'en bloc' groin node dissection may be warranted *(Hacker et al., 1981)*.

Reconstructive surgical techniques may be appropriate to enable primary surgical closure and reduce morbidity due to scarring and may involve colorectal, and plastic surgeons.
Primary radiotherapy or chemoradiotherapy

These can be considered for selected patients with advanced disease such as those who are not suitable for surgery because of the extent of their disease or co-existing medical conditions. However, caution is needed when dealing with frail, often elderly, patients in view of the potential toxicity of treatment.

**Table 2 Summary of primary treatment of squamous vulval cancer**

<table>
<thead>
<tr>
<th>Disease State</th>
<th>Description</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early disease</td>
<td>Small lesions with less than 1mm invasion (FIGO Stage IA)</td>
<td>Wide local excision, groin node dissection unnecessary</td>
</tr>
<tr>
<td></td>
<td>Truly lateralised squamous lesions (FIGO Stage IB and II)</td>
<td>Initially only require wide local excision and ipsilateral lymphadenectomy</td>
</tr>
<tr>
<td></td>
<td>Centrally located tumours where excision is possible without sphincter compromise</td>
<td>Requires wide local excision and bilateral lymphadenectomy initially</td>
</tr>
<tr>
<td>Advanced disease</td>
<td>Extensive vulval involvement</td>
<td>Groin node dissection and either surgical excision (consider vulval reconstruction) or primary (chemo)radiotherapy</td>
</tr>
<tr>
<td></td>
<td>Clinically advanced nodes</td>
<td>Excision and/or chemo radiation therapy</td>
</tr>
<tr>
<td>Metastatic disease</td>
<td></td>
<td>Palliation may require surgical management of the primary tumour</td>
</tr>
</tbody>
</table>

**8.3. Surgical management of non squamous vulval cancer**

**Carcinoma of the Bartholin’s gland**

This is rare and can be either a squamous carcinoma or adenocarcinoma histologically. The current evidence base is insufficient to suggest different management from squamous tumours.
Basal cell carcinoma and verrucous carcinoma

These squamous variants are rarely associated with lymph node metastases and can be managed by wide local excision. Basal cell carcinomas are also amenable to treatment by radiotherapy, which should be the preferred treatment if resection would compromise function (for example anal sphincter damage).

Malignant melanomas

In cases of suspected melanoma one should aim for an initial excision of the whole lesion with a 2mm margin where possible. This allows for histopathological examination of the whole lesion for assessment of the Breslow thickness (Breslow, 1970). Depending on the clinical situation, it may sometimes be necessary to take an initial biopsy first.

Once the diagnosis of melanoma has been established then the case should immediately be discussed with members of the Local Skin Cancer MDT, for guidance on the margin of wide local excision and whether the patient may be considered for sentinel node biopsy at the time of wide local excision.

If there is clinically apparent lymphadenopathy at the time of presentation, then the case should be discussed with the Regional Skin Cancer MDT, regarding initial staging investigations which may be required prior to definitive surgery, and also the extent of the definitive surgery

Sarcoma of the vulva

Sarcoma of the vulva is rare and has a tendency to local blood borne spread. Surgery is often combined with radiotherapy and chemotherapy. These cases should be discussed with the sarcoma MDT.
9. Management of recurrent disease

Twenty four per cent of patients develop recurrent disease, which is usually on the vulva (Salmon, 2002).

9.1. Local recurrence

Local recurrence is managed by repeat excision, especially if irradiation has already been given to maximum dose. If excision will impair sphincter function then radiotherapy should be considered. Plastic surgery with rotational skin and myocutaneous flaps may be necessary. The patient should be referred to the Cancer Centre for appropriate assessment by the multidisciplinary team.

9.2. Regional recurrence

Groin recurrence is much more difficult to manage. In patients who have not received radiation this should be performed first, followed by resection if the response to radiotherapy is not complete. Patients who have been irradiated should be offered palliative resection if possible. Survival after groin recurrence is poor. The palliative care team should be involved early (Royal College of Obstetrician and Gynaecologists, 2006).

9.3. Chemotherapy and radiotherapy for recurrent disease

For selected patients with loco-regional recurrences who have been treated with surgery alone, radiotherapy or chemoradiotherapy can be considered and may offer the prospect of disease control (Thomas et al., 1989). For patients with recurrent and progressive vulval cancer, palliative chemotherapy may be appropriate in some cases, although responses are often short-lived. Frequent complications from the cancer include infection, bleeding and lymphoedema, and chemotherapy should be given in the context of multi-disciplinary palliative care.
10. **Vulval cancer - consideration of aetiology-specific management**

Vulval cancer is rare and the suggestions in this section are based on expert opinion, rather than evidence based guidelines.

Documentation of the clinical aetiology of vulval cancer will allow appropriate management. If there is a history of HPV-related disease (for example Cervical Intraepithelial Neoplasia [CIN]), or the appearance is suggestive (discrete lesion, no evidence of lichen sclerosus), then consideration should be given to regular examination of the vagina, perianal skin and increased frequency of cervical smears. If there is evidence of lichen sclerosus, then this should be controlled pre-operatively. Surgery often causes lichen sclerosus to flare up. Very potent steroids (for example clobetasol propionate ointment) and information on care of the vulva (for example avoiding all vulval contact with soap) are appropriate to keep the disease controlled both pre and post operatively.

11. **Vulval reconstruction**

Vulval cancer is rare and the following section contains suggestions based on expert opinion, rather than evidence based guidelines.

Reconstruction following vulvectomy may improve function post operatively, but is still associated with significant psychosexual morbidity. Guidelines for reconstruction associated with breast cancer recommend the pre-operative use of images to show women what to expect. Although this may seem difficult, women do appear to benefit from a realistic idea of what to expect post-operatively. Reconstructive techniques include sliding V-Y flaps and rotational gluteal fold flaps. For the latter, the input of a plastic or reconstructive surgeon is recommended. Split thickness skin grafts are generally not appropriate for use in the vulva, as the area does not lend itself to optimal healing and the shrinkage that occurs can leave troublesome scarring and stenosis.
12. Follow up

Follow up intervals are currently arbitrary, but the following schedule is suggested:

- Three monthly first year.
- Six monthly second year.
- Annual review for 5-10 years.

13. Lymphoedema

All women undergoing groin node dissection should be referred to a specialist lymphoedema service prior to surgery if possible to educate patients and establish baseline circumference measurements of the lower limbs. Lymphoedema education consists of skin care advice, exercise, Simple Lymphatic Drainage massage and possibly compression garments. All patients undergoing groin node dissection, or sentinel node biopsy only, should be followed up as outpatients in the lymphoedema service once discharged from hospital. Proactive treatment can significantly reduce lymphoedema and control swelling even in the presence of progressive disease. The Clinical Nurse Specialist (CNS) will also reinforce lymphoedema education and give advice as appropriate.

All women who develop lower-limb lymphoedema should have access to the four cornerstones of lymphoedema care;

- skin care to maintain a good tissue condition and reduce the risk of infection
- external compression in the form of elastic compression garments that help reduce new lymph formation and encourage lymph drainage by improving the efficiency of muscle pump
- a programme of exercise and movement to promote lymph drainage without over exertion
- simple lymphatic drainage: a method of lymph drainage that can be carried out by the patient or carer and involves a series of simple hand movements.
The aims of this regimen are to rehabilitate the cancer patient, to reduce any disability as far as possible, to help the patient to achieve an independent lifestyle and to give the patient the skills to manage their own condition (British Lymphology Society, 2009)

14. The Clinical Nurse Specialist (CNS) role

All women with a diagnosis, of a vulval cancer should be offered the support of, and have access to, a CNS in order to facilitate the woman’s needs throughout the cancer journey, including those of her partner or carer.

Within an MDT, the CNS is an ideal position, frequently as the key worker, to be able to address the often complex and sensitive issues identified and experienced by the patient (NICE, 2004).

A holistic needs assessment should be carried out upon diagnosis and the individualised patient issues/concerns, such as altered body image and sexual dysfunction, should be addressed by appropriately trained specialist nurses, counselors or clinical psychologists (Macmillan, 2013).

The use of clear, accurate written information and photographs/drawings should be promoted and given to patients and with their consent, their partners (Corney et al., 1992; Macmillan, 2013).

Access to self-help and support groups, such as the Vulval Awareness Campaign Organisation, may also be of significant benefit, allowing women to share experiences and seek support from other women diagnosed and treated for the same condition.
All sexually active patients and their partners should be offered specific information on the effect of treatment; surgery and radiotherapy, on their relationship and be advised of the specialist role of the CNS (National Health Service Executive, 1999). The advanced communication skills, expertise and confidence in discussion of sexuality issues, which are encompassed within the CNS role suggests this route is most advantageous.
15. Example flow chart for the management of vulval tumours
16. References


British Lymphology Society. Strategy for lymphoedema care February 2009


Royal College of Obstetricians and Gynaecologists. Guidelines for the Diagnosis and Management of Vulval Carcinoma. May 2014

Royal College of Obstetrician and Gynaecologists, Management of vulva cancer. 2006

Royal College of Pathologists. Datasets for the histopathological reporting of vulval neoplasms 3rd edition 2010


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Appendix 1 Guideline methodology.

The Cancer NSAG Gynaecology sub-group responded to Cancer Network requests to update the “Management of Gynaecological Cancers 2001” guideline. Agreement was reached to review each disease separately starting with vulval cancer.

A multidisciplinary subgroup was formed, led by Amanda Tristam as lead author, with representatives from each 3 regions in Wales and the following steps undertaken:

- Literature sourcing
- Review of evidence using Oxford 2011 (see Appendix 2)
- Iterative drafting of vulval guideline
- Presentation to Cancer NSAG Gynaecology subgroup for comment.
- Review post comments from NSAG
- Editorial sweep by Cancer NSAG Core Team
- Sign-off for consultation by Cancer NSAG Gynaecology subgroup
- 1 month consultation to stakeholders including:
  - all gynaecology MDT members
  - cervical screening
  - Cancer NSAG skin subgroup
  - Cancer NSAG colorectal subgroup
  - Cancer Networks
  - Cancer Executive leads for all LHBs and Velindre NHS Trust
- editorial group to respond to consultation comments and amend doc as necessary
- sign off to publish by Cancer NSAG
- circulation to Welsh Medical Committee for agreement

To follow
- publication by Welsh Medical Committee
# Appendix 2 Oxford Centre for Evidence-Based Medicine 2011 Levels of Evidence.

<table>
<thead>
<tr>
<th>Question</th>
<th>Step 1 (Level 1*)</th>
<th>Step 2 (Level 2*)</th>
<th>Step 3 (Level 3*)</th>
<th>Step 4 (Level 4*)</th>
<th>Step 5 (Level 5)</th>
</tr>
</thead>
<tbody>
<tr>
<td>How common is the problem?</td>
<td>Local and current random sample surveys (or censuses)</td>
<td>Systematic review of surveys that allow matching to local circumstances**</td>
<td>Local non-random sample**</td>
<td>Case-series**</td>
<td>n/a</td>
</tr>
<tr>
<td>Is this diagnostic or monitoring test accurate?</td>
<td>Systematic review of cross sectional studies with consistently applied reference standard and blinding</td>
<td>Individual cross sectional studies with consistently applied reference standard and blinding</td>
<td>Non-consecutive studies, or studies without consistently applied reference standard**</td>
<td>Case-control studies, or poor or non-independent reference standard**</td>
<td>Mechanism-based reasoning</td>
</tr>
<tr>
<td>What will happen if we do not add a therapy?</td>
<td>Systematic review of inception cohort studies</td>
<td>Inception cohort studies</td>
<td>Cohort study or control arm of randomized trial*</td>
<td>Case-series or case-control studies, or poor quality prognostic cohort study**</td>
<td>n/a</td>
</tr>
<tr>
<td>Does this intervention help?</td>
<td>Systematic review of randomized trials or n-of-1 trials</td>
<td>Randomized trial or observational study with dramatic effect</td>
<td>Non-randomized controlled cohort/follow-up study**</td>
<td>Case-series, case-control studies, or historically controlled studies**</td>
<td>Mechanism-based reasoning</td>
</tr>
<tr>
<td>What are the COMMON harms?</td>
<td>Systematic review of randomized trials, systematic review of nested case-control studies, n-of-1 trial with the patient you are raising the question about, or observational study with dramatic effect</td>
<td>Individual randomized trial or (exceptionally) observational study with dramatic effect</td>
<td>Non-randomized controlled cohort/follow-up study (post-marketing surveillance) provided there are sufficient numbers to rule out a common harm. (For long-term harms the duration of follow-up must be sufficient.)**</td>
<td>Case-series, case-control, or historically controlled studies**</td>
<td>Mechanism-based reasoning</td>
</tr>
<tr>
<td>What are the RARE harms?</td>
<td>Systematic review of randomized trials or n-of-1 trial</td>
<td>Randomized trial or (exceptionally) observational study with dramatic effect</td>
<td>Non-randomized controlled cohort/follow-up study**</td>
<td>Case-series, case-control, or historically controlled studies**</td>
<td>Mechanism-based reasoning</td>
</tr>
<tr>
<td>Is this (early detection) test worthwhile?</td>
<td>Systematic review of randomized trials</td>
<td>Randomized trial</td>
<td>Non-randomized controlled cohort/follow-up study**</td>
<td>Case-series, case-control, or historically controlled studies**</td>
<td>Mechanism-based reasoning</td>
</tr>
</tbody>
</table>

* Level may be graded down on the basis of study quality, imprecision, indirectness (study PICO does not match questions PICO), because of inconsistency between studies, or because the absolute effect size is very small; Level may be graded up if there is a large or very large effect size.

** As always, a systematic review is generally better than an individual study.