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MAIN TEXT

VULVAL LICHEN SCLEROSUS: AN AUSTRALASIAN MANAGEMENT CONSENSUS

INTRODUCTION

Vulval lichen sclerosus (VLS) is a chronic inflammatory skin condition predominantly affecting the anogenital region in women and children. The prevalence is unknown due to underdiagnosis and referral bias, however it has been reported to account for at least 10% of new cases in vulval practice(1). Symptoms of VLS include pruritus, pain, and dyspareunia and this may severely impact the patient's quality of life and sexual functioning(2). If neglected or inadequately treated, VLS may lead to loss of architecture due to scarring with loss of labia, burying of the clitoris and stenosis of the vaginal introitus(3, 4). VLS is also associated with a 3.5% – 6% lifetime risk of developing vulval squamous cell carcinoma (SCC)(5, 6). VLS can be asymptomatic and subsequently discovered during gynaecological examinations for unrelated issues. Unfortunately, it may also present at an advanced stage of the disease or when carcinoma has arisen(7).

There are no existing guidelines for the management of VLS in the Australasian population. Guidelines to achieve and maintain remission and the ideal duration of follow-up for VLS vary(8, 9). Some guidelines suggest women with problematic VLS be followed up regularly by specialists, and patients with uncomplicated disease be discharged to their primary care physician(8, 10). In contrast, an Australian study on the long-term management of adult VLS recommended regular long-term surveillance for all patients(11). Actual practice may be

affected by a variety of factors such as the availability or scarcity of individual specialists and/or specialist vulval clinics.

The aim of this study is to document a consensus approach to VLS management in the Australasian setting.

Key practice points

- It is recommended that a biopsy or photographic records be taken at first presentation and/or prior to treatment in all patients.
- Potent to ultra-potent topical corticosteroids are the mainstay of initial treatment for VLS.
- Long-term treatment should be adjusted according to the severity of disease.
- Long-term individualised regimens with topical corticosteroids (TCS) are recommended to achieve optimal outcomes.
- Continue regular TCS treatment even when asymptomatic.
- Follow up of VLS should be undertaken for an indefinite period.

MATERIALS AND METHODS

An initial focus group of 12 medical practitioners with expertise in VLS, consisting of dermatologists, gynaecologists and a paediatrician, were invited to share their clinical expertise and develop a draft consensus statement for the severity scoring and management of vulval lichen sclerosus.

Study type

The online Delphi (eDelphi) method is an established, iterative, multi-stage process for developing consensus guidelines. It is widely used in clinical services and health research for establishing diagnostic criteria and management guidelines(12). A panel of experts in the relevant field are invited to vote anonymously on a series of statements using a web-based interface. After each subsequent visit, a summary of responses is revealed to the panel. Participants are then given the opportunity to revise their opinion or keep their original answers for the next round, while taking into consideration the group's response.

Participants

Recruitment to the panel for the eDelphi study consisted of targeted invitations to those already known to the focus group who had relevant clinical expertise in the field of VLS. A panel of 22 members from Australia and New Zealand comprised 15 dermatologists, six gynaecologists, and one paediatrician. The panel then voted on the draft consensus statement developed by the focus group, using a modified eDelphi process.

Consent to participate was implied if individual members completed each round, as specified in the invitation letter for the consensus study.

Study procedures

The eDelphi consensus study was carried out between April to August, 2020. It was moderated by a single coordinator, who did not participate in the survey rounds. The consensus process included four rounds of remote voting in which panel members were asked to anonymously rate statements related to the diagnosis, severity, initial and long-term management, follow-up and complications of VLS.

For each round, participants were asked to state their agreement with each statement using a 5-point Likert-scale, from 1 (strongly disagree) to 5 (strongly agree). Participants were also encouraged to provide additional free-text comments to generate further statements for inclusion. In round 1, demographic data about the participants was also collected. After round 1 of voting, the responses were collected and analysed. A summary for each statement was provided to the participants, including the distribution of the group scores (level of consensus), the strength score (median score), and their individual score.

In round 2, participants were asked to state their agreement with rephrased statements for which consensus was not met, for which wording clarification was requested in round 1, and for additional items suggested in round 1. A summarised report of the findings was again distributed to each participant, along with a reminder of the score they gave for each statement in round 2. Likewise, revised statements were presented to participants in rounds 3 and 4.

Definition of consensus

The definition of 'consensus' used for this study was in accordance with previous literature(13, 14). Consensus was also defined as a priori when 70% or more of participants were in agreement (score of 4–5; agree to strongly agree) or were in disagreement (1–2; disagree to strongly disagree).

All round 1 participants were invited to subsequent rounds and given a maximum of two weeks to submit their responses. To address a potential attrition bias, participants were reminded to complete each survey at the end of each round. A maximum of three reminders were sent to participants who agreed to partake but had not completed the survey.

RESULTS

Participants' characteristics

Twenty-two members of the panel completed all four rounds of the survey. The panel members were highly experienced VLS specialists who predominately each reviewed more than 100 patients annually. The panel members were also representative of a range of specialities, as well as major geographic regions across Australia and New Zealand (Table 1).

Response rates

Members of the panel were encouraged to vote on all statements at each round. A high response rate was seen for all statements at each round. In the first round, 14 participants submitted their response to all statements and eight members did not respond to at least one statement. In the second round, three participants declined to respond to at least one statement. In rounds 3 and 4, all 22 participants responded to all statements.

By the end of round 1, 37 out of 48 statements reached consensus. Items in round 2 were fewer and included four new statements generated from round 1. Of the 11 statements from the first round, six were modified and five items were repeated. By round 3, three statements reached consensus and one statement was removed. By the final round of voting, there was agreement on a total of 51 statements supporting the proposed management guideline of vulval lichen sclerosus (Tables 2–5). Additional commentary resulted in minor editing.

DISCUSSION

This eDelphi study was used to gain consensus for best practice management of VLS in an Australasian setting. The panel agreed on a core set of guideline statements related to the diagnosis, severity, initial and long-term management, follow-up and complications of VLS which were used to construct an algorithm for adult patients with VLS (Fig.1).

The eDelphi method(15) involved a panel of experts and multiple iterations, feedback after each round, and quasi anonymity. The commitment of the 22 experts throughout the 4-month study period, indicated their interest in improving patient care for VLS. The distribution of the panel members was a fair representation of the geographical Australasian setting, with each member having the necessary skills and experience to contribute to the consensus guideline, thus strengthening their recommendations.

Diagnosis and severity of patients with VLS

The panel agreed unanimously in the value of a biopsy or photographic records in adults to aid diagnosis prior to treatment. The majority agreed that the use of photographs is desirable to monitor progress. A subsequent biopsy is recommended if malignancy is suspected or when there is uncertainty regarding the diagnosis, or in the patient who is unresponsive to first line treatment despite appropriate intervention. The panel mostly agreed that VLS in children should be considered a clinical diagnosis, although all members recommended that biopsy should be conducted if there is doubt in the diagnosis, or if the disease fails to respond to adequate treatment.

While there has been little agreement on a VLS severity scoring system to date, the panel affirmed that severity scoring should be based on physician assessment and supplemented with a Quality of Life assessment tool such as the VQLI (Vulval Quality of life Index)(16). VLS has a significant impact on the quality of life, including sexual function(2).

Initial management

Previous management guidelines for VLS originating from the UK, Europe and the USA support the use of 0.05% clobetasol propionate ointment(4, 8, 17). Studies have also shown other potent and ultra-potent TCS to be effective and safe in treating adults and

children(18). Clobetasol propionate 0.05% ointment is not easily available in Australia and although the panel mostly agreed that potent to ultra-potent TCS should be the first line of treatment for inducing remission in VLS it did not specifically support clobetasol propionate 0.05% as standard of care. A standard therapy was not specified in this study as treatment choice should be individualised and adjusted according to disease severity. For most Australasian practitioners other potent or super-potent preparations have also proved to be effective as initial treatment. For example, in previously published studies, mometasone furoate 0.1% has been shown to be effective in treating VLS(18-20).

Few publications discuss VLS during pregnancy. A recent case series showed that treatment with TCS is safe during pregnancy, without requiring adjustment from the non-pregnant state(21). It was agreed among the majority of panel members that the same TCS treatment should be used for women with VLS who are pregnant.

Long term management

VLS is a lifelong disease that, without treatment, can lead to the loss of structure of the vulva(22). Patients who remain asymptomatic are still at risk of complications including carcinoma(7). A recently published study has shown that long-term treatment greatly improves quality of life and reduces risk of scarring (JLGTD in press). In contrast to European and British guidelines, which recommend treating only active LS during maintenance therapy(4, 8), an Australian study has shown that regular, long term, preventative treatment with TCS that achieve objective normality of skin leads to better disease outcomes(22). The same study also established that 'as needed' treatment for symptom control may not reduce the risk of developing cancer(22). While a number of reviews (17, 23, 24) also recommend treatment based on symptom control, the majority on the panel agreed that intermittent, or 'as required' TCS, is not recommended and agreed that patients should use preventative TCS treatment regularly despite being asymptomatic.

Recent studies have reported that individualised long term TCS treatment in a compliant population, improves function, relieves symptoms, and prevents the progression of scarring and carcinoma of the vulva(22, 25). Most panel members agreed that long-term treatment with individualised regimes with TCS is recommended. If improved, treatment should be

maintained to ensure compliance, adjust and maintain treatment regime, and monitor for complications with indefinite follow-up. If not improved and the patient relapses in the absence of another condition, TCS potency and/or frequency of application should be increased.

Follow-up

Given individual variability and the time required for individuals to achieve therapeutic response, the panel agreed that the initial follow-up appointment should be no longer than 12 weeks and those with unstable or complicated disease should be reviewed earlier. It was agreed that this cohort should be assessed every three months, whereas people with stable or uncomplicated disease can be assessed and monitored every 6–12 months. In patients who have had VLS associated vulval SCC/differentiated vulval intraepithelial neoplasia (dVIN) follow up should remain under specialist care at appropriate intervals determined by the specialist.

Current UK and European management guidelines recommend discharging patients with stable uncomplicated disease back to their primary care physician(4, 8). However a significant portion of patients are not seen by their primary care physician for follow-up, and if reviewed, not examined during their follow-up visit(26). The panel agreed that for stable, well controlled uncomplicated disease, follow-up of patient can be managed by a primary physician provided there is shared care with a review by a specialist at least every 2 years.

Calcineurin inhibitors and LS

A recent systematic review found that topical pimecromilus was less effective than clobetasol propionate in improving gross appearance and reducing inflammation(18, 27). Further, there have been case reports of vulval cancer in LS patients treated with pimecrolimus(28).The panel members agreed that topical calcineurin inhibitors (TCIs) such as pimecromilus do not offer any advantage over TCS with treatment resistant VLS. The safety profile of TCS in the long term management of VLS has been established in reported studies(22, 29). It was agreed among the majority of panel members that the long term use of TCS in patients with VLS is safe with minimal adverse events, which are treatable and mostly reversible.

Limitation

A limitation is that the panel did not include patients; clinical decisions for patients with VLS are based on disease severity as determined by the healthcare provider.

Conclusion

The eDelphi exercise has resulted in strong consensus on a set of approaches crucial to the diagnosis and short and long-term management of VLS. This guideline was devised by Australasian experts however we hope that it will be generalisable to patients with LS worldwide.

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Table 1. Characteristics of the Delphi panel members

Characteristics	Participants (n= 22)	%
Occupation		
Dermatologist	16	72.7
Gynaecologist	5	22.7
Paediatrician	1	4.5
Number of patients (new/old) with VLS in a year		
10-25	1	4.5
26-50	1	4.5
51-75	3	13.6
76-100	2	9.1
>100	15	68.2
Principal region of practice		
Victoria	6	27.3
New South Wales (incl. Australian Capital Territory)	5	22.7
South Australia	4	18.2
New Zealand	3	13.6
Western Australia	2	9.1
Tasmania	1	4.5
Queensland	1	4.5

Experience with managing VLS, years		
<10	1	4.5
10-19	5	22.7
20-30	11	50.0
>30	5	22.7

Table 2. Severity and Diagnosis of VLS

Consensus recommendations	Strength	Consensus
Severity scoring should be based on physician assessment rather than patient reported symptoms	4	95%
When assessing the severity of VLS, consider the following signs on examination:		

[1] Fusion (Agglutination)*	4	82%
[2] Lichenification	4	71%
[3] Loss of vulval architecture (e.g. clitoral hood fusion, labial fusion, narrowing of the introitus)**	4	91%
[4] Extent of disease (e.g. small area vs. entire genital, perineum, perianal skin)	4	86%
[5] Ulceration	4	95%
[6] Purpura	4	81%
Severity items should be measured on a scale of none (0), mild (1), moderate (2), severe (3) or very severe (4)	4	95%
A supplemental QoL assessment tool [such as the VLQI (vulval lichen quality index) tool] should be used when assessing severity of patient reported symptoms and to monitor improvement	4	82%
Diagnosis of VLS		
Ideally, a biopsy or photographic records in adults should be taken at first presentation and/or prior to treatment.	5	100%
When symptoms are disproportionate to physician assessed severity, one must consider other reasons for discomfort eg neuropathy, candida etc.	5	95%
If a patient with objectively well controlled disease on examination is symptomatic, the symptoms may not be related to the VLS itself	4	95%
Photographic records are desirable in monitoring progress	5	91%
Failure to improve with adequate TCS should be a reason to suspect an alternative diagnosis	4	100%
Patients may require a subsequent biopsy when		
- The clinical diagnosis is in doubt	4	86%
- If recommended first line treatment fails after appropriate treatment duration	4	82%
- If malignancies are suspected	5	91%
Diagnosis of VLS is compatible with a normal vaginal birth	5	95%
Diagnosis of childhood LS should be on clinical grounds	4	95%
Biopsy in children should be reserved only if there is doubt in diagnoses or resistance to adequate treatment	4	100%

**Fusion: loss of definition between structures due to agglutination of surfaces.*

*** Loss of vulval architecture: Missing structures such as labia minora, visible clitoral tip, buried clitoris.*

Table 3. Initial Management

Consensus recommendations	Strength	Consensus
Initial treatment (first 6 months) outcome of VLS should		
- Relieve symptoms	5	95%
- Induce remission: attaining normal or near to normal skin colour and texture (Note: there may be residual hyper-pigmentation or pallor)	4	91%
Potent to ultra-potent topical corticosteroids are the mainstay of initial treatment for VLS.	5	95%
Patients should be counselled on long term consequences of VLS, specifically,		
- Risk of squamous cell carcinoma of the vulva	5	91%
- Scarring/loss of structure is permanent and not changed by treatment	5	90%
- Risk of recurrence if treatment ceased or inadequate	5	100%
Patients should be educated on the chronic nature of LS, which requires ongoing treatment to keep under control	5	100%
Treatment (e.g. TCS potency/frequency of application) should be adjusted according to severity of disease	5	95%
First follow up should be no longer than 12 weeks, depending on severity	4.5	86%
TCS treatment regime in pregnancy should be the same as in the non-pregnant woman	4.5	91%
Treatment of LS in children should be supervised by a specialist	5	100%
Maintenance Management		
The treatment outcome during maintenance is to provide		
- Symptom control	5	95%
- Prevention of scarring	4	91%
- Lack of side effects	4	82%
- Reduction of carcinoma risk	4	86%
- Objective disease suppression, maintenance of normal or near to normal skin colour and texture.	4	86%
The desired long-term treatment with individualised regimens with TCS to achieve the treatment outcomes is recommended	5	100%
Patients should use preventative TCS treatment regularly despite being asymptomatic.	5	81%
A treatment regimen using intermittent or 'as required' TCS to control symptoms only is NOT recommended.	5	82%
Once a patient has improved, treatment should be maintained provided there are no side effects and the condition is well controlled	4	91%
During maintenance period, if a patient relapses in the absence of another condition TCS potency and/or frequency of application should be increased.	5	100%

During maintenance period, if a patient experiences adverse effects from TCS such as atrophy or corticosteroid dermatitis (provided patient has been properly counselled on application and candidiasis has been excluded) treatment should be adjusted accordingly (i.e potency and/or frequency).	5	100%

Table 4. Follow up

Consensus recommendations	Strength	Consensus
Follow up of LS should be for an indefinite period	4.5	95%
When managing patients with VLS, regular follow-up is recommended to optimise compliance, adjust and maintain treatment regime, and monitor for complications	5	95%
For stable uncomplicated disease, patients should be reassessed every 6-12 months	4	91%
For unstable complicated disease, patients should be reassessed least every 3 months, depending on the individual and/or severity.	4	96%
For stable, well controlled uncomplicated disease, follow-up of patient can be managed by a primary physician provided there is shared care with a review by a specialist at least every 2 years.	4	73%

Table 5. Managing Complications

Consensus recommendations	Strength	Consensus
The management of treatment resistant VLS in adults and children		
- Should be undertaken by a specialist experienced in managing complex cases of VLS	4	81%
- Topical calcineurin inhibitors (TCIs) do not offer any advantage over TCS	4	77%
The use of long-term TCS in patients (adults and children) with VLS is safe with minimal adverse events.	5	95%
Adverse events related to TCS use in patients with VLS such as atrophy, stinging, erythema, allergic contact dermatitis, TCS induced dermatitis and superinfection are treatable and/or reversible.	4	95%

Figure 1. Recommended management pathway for Adult Vulval lichen sclerosus

Table 1. Characteristics of the Delphi panel members

Characteristics	Participants (n= 22)	%
Occupation		
Dermatologist	16	72.7
Gynaecologist	5	22.7
Paediatrician	1	4.5
Number of patients (new/old) with VLS in a year		
10-25	1	4.5
26-50	1	4.5
51-75	3	13.6
76-100	2	9.1
>100	15	68.2
Principal region of practice		
Victoria	6	27.3
New South Wales (incl. Australian Capital Territory)	5	22.7
South Australia	4	18.2
New Zealand	3	13.6
Western Australia	2	9.1
Tasmania	1	4.5
Queensland	1	4.5
Experience with managing VLS, years		
<10	1	4.5
10-19	5	22.7
20-30	11	50.0
>30	5	22.7

Table 2. Severity and Diagnosis of VLS

Consensus recommendations	Strength	Consensus
Severity scoring should be based on physician assessment rather than patient reported symptoms	4	95%
When assessing the severity of VLS, consider the following signs on examination:		
[1] Fusion (Agglutination)*	4	82%
[2] Lichenification	4	71%
[3] Loss of vulval architecture (e.g. clitoral hood fusion, labial fusion, narrowing of the introitus)**	4	91%
[4] Extent of disease (e.g. small area vs. entire genital, perineum, perianal skin)	4	86%
[5] Ulceration	4	95%
[6] Purpura	4	81%
Severity items should be measured on a scale of none (0), mild (1), moderate (2), severe (3) or very severe (4)	4	95%
A supplemental QoL assessment tool [such as the VLQI (vulval lichen quality index) tool] should be used when assessing severity of patient reported symptoms and to monitor improvement	4	82%
Diagnosis of VLS		
Ideally, a biopsy or photographic records in adults should be taken at first presentation and/or prior to treatment.	5	100%

When symptoms are disproportionate to physician assessed severity, one must consider other reasons for discomfort eg neuropathy, candida etc.	5	95%
If a patient with objectively well controlled disease on examination is symptomatic, the symptoms may not be related to the VLS itself	4	95%
Photographic records are desirable in monitoring progress	5	91%
Failure to improve with adequate TCS should be a reason to suspect an alternative diagnosis	4	100%
Patients may require a subsequent biopsy when		
- The clinical diagnosis is in doubt	4	86%
- If recommended first line treatment fails after appropriate treatment duration	4	82%
- If malignancies are suspected	5	91%
Diagnosis of VLS is compatible with a normal vaginal birth	5	95%
Diagnosis of childhood LS should be on clinical grounds	4	95%
Biopsy in children should be reserved only if there is doubt in diagnoses or resistance to adequate treatment	4	100%

**Fusion: loss of definition between structures due to agglutination of surfaces.*

*** Loss of vulval architecture: Missing structures such as labia minora, visible clitoral tip, buried clitoris.*

Table 3. Initial and Maintenance Management of VLS

Consensus recommendations	Strength	Consensus
Initial treatment (first 6 months) outcome of VLS should		
- Relieve symptoms	5	95%
- Induce remission: attaining normal or near to normal skin colour and texture (Note: there may be residual hyper-pigmentation or pallor)	4	91%

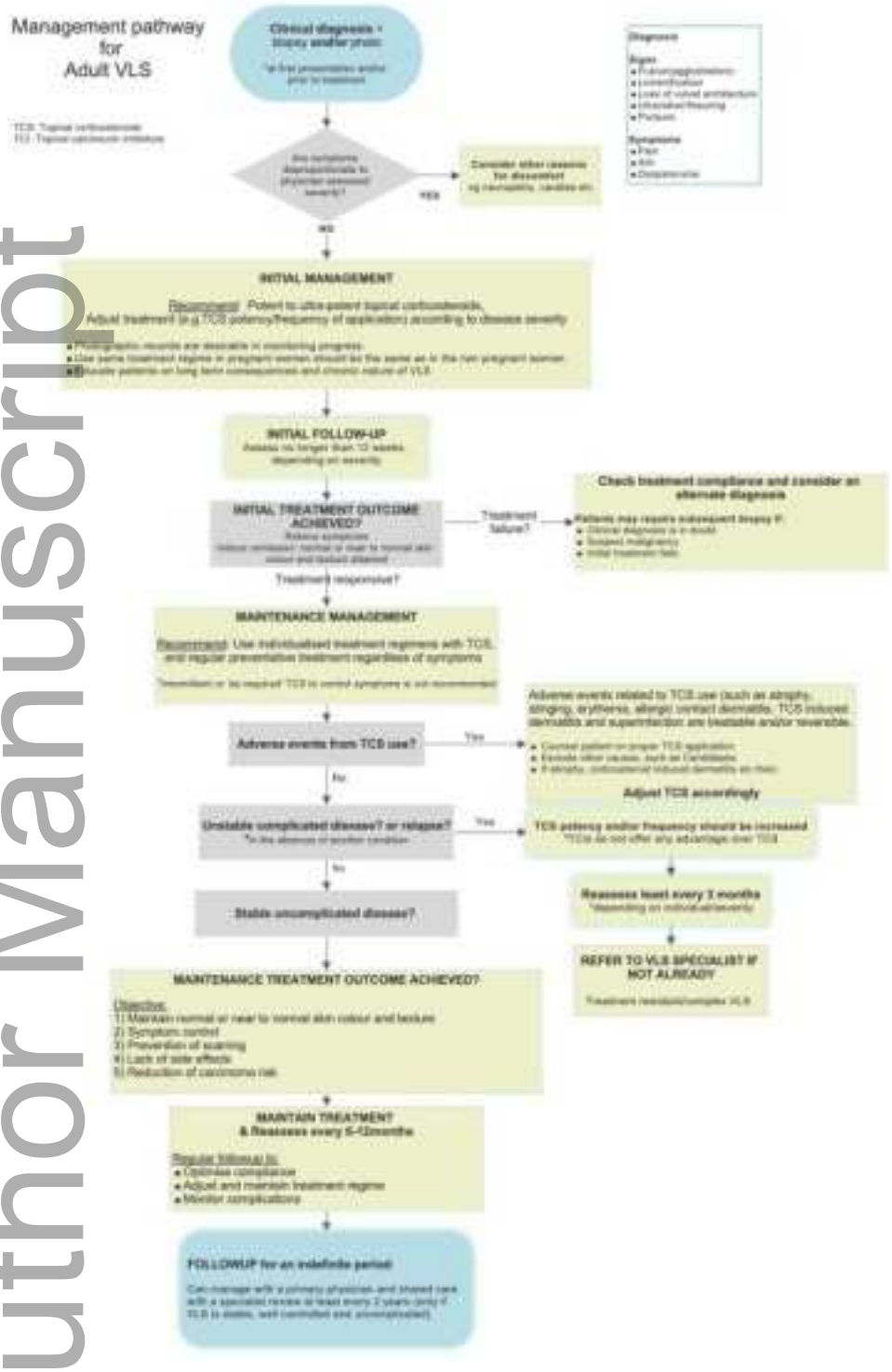
Potent to ultra-potent topical corticosteroids are the mainstay of initial treatment for VLS.	5	95%
Patients should be counselled on long term consequences of VLS, specifically,		
- Risk of squamous cell carcinoma of the vulva	5	91%
- Scarring/loss of structure is permanent and not changed by treatment	5	90%
- Risk of recurrence if treatment ceased or inadequate	5	100%
Patients should be educated on the chronic nature of LS, which requires ongoing treatment to keep under control	5	100%
Treatment (e.g. TCS potency/frequency of application) should be adjusted according to severity of disease	5	95%
First follow up should be no longer than 12 weeks, depending on severity	4.5	86%
TCS treatment regime in pregnancy should be the same as in the non-pregnant woman	4.5	91%
Treatment of LS in children should be supervised by a specialist	5	100%
Maintenance Management		
The treatment outcome during maintenance is to provide		
- Symptom control	5	95%
- Prevention of scarring	4	91%
- Lack of side effects	4	82%
- Reduction of carcinoma risk	4	86%
- Objective disease suppression, maintenance of normal or near to normal skin colour and texture.	4	86%
The desired long-term treatment with individualised regimens with TCS to achieve the treatment outcomes is recommended	5	100%
Patients should use preventative TCS treatment regularly despite being asymptomatic.	5	81%
A treatment regimen using intermittent or 'as required' TCS to control symptoms only is NOT recommended.	5	82%
Once a patient has improved, treatment should be maintained provided there are no side effects and the condition is well controlled	4	91%
During maintenance period, if a patient relapses in the absence of another condition TCS potency and/or frequency of application should be increased.	5	100%
During maintenance period, if a patient experiences adverse effects from TCS such as atrophy or corticosteroid dermatitis (provided patient has been properly counselled on application and candidiasis has been excluded) treatment should be adjusted accordingly (i.e potency and/or frequency).	5	100%

Table 4. Follow up of VLS

Consensus recommendations	Strength	Consensus
Follow up of LS should be for an indefinite period	4.5	95%
When managing patients with VLS, regular follow-up is recommended to optimise compliance, adjust and maintain treatment regime, and monitor for complications	5	95%
For stable uncomplicated disease, patients should be reassessed every 6-12 months	4	91%
For unstable complicated disease, patients should be reassessed least every 3 months, depending on the individual and/or severity.	4	96%
For stable, well controlled uncomplicated disease, follow-up of patient can be managed by a primary physician provided there is shared care with a review by a specialist at least every 2 years.	4	73%

Table 5. Managing Complications

Consensus recommendations	Strength	Consensus
The management of treatment resistant VLS in adults and children		
- Should be undertaken by a specialist experienced in managing complex cases of VLS	4	81%
- Topical calcineurin inhibitors (TCIs) do not offer any advantage over TCS	4	77%
The use of long-term TCS in patients (adults and children) with VLS is safe with minimal adverse events.	5	95%
Adverse events related to TCS use in patients with VLS such as atrophy, stinging, erythema, allergic contact dermatitis, TCS induced dermatitis and superinfection are treatable and/or reversible.	4	95%



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