Vulval conditions may present with a rash but no other symptoms or with symptoms but no significant rash. Causes of the former include tumours, cysts and warts; causes of the latter include referred and neuropathic pain and atrophic vaginitis.

Vulval disease

Part 3: patients with a rash, a lesion, or other symptoms, alone

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In Part 3 of this three-part article about vulval disease, we describe those conditions that present with either a rash or other symptoms but not both. Part 1 presented the principles of a dermatogynaecological approach to the evaluation, investigations and management of vulval conditions; Part 2 described those conditions that present with rash and other symptoms.

The patient with a rash or lesion alone

Asymptomatic vulval lesions may be noticed by the patient or patient's partner or discovered on routine Pap smear; they sometimes require investigation and treatment. The Table lists causes of a vulval rash or lesion without other symptoms. VIN III and lichen sclerosus may present with a rash or lesion alone or with other symptoms and have been discussed in Part 2 of this article. Here, we look at other causes of a vulval rash or lesion in the absence of other symptoms: melanosis vulvae, benign and malignant tumours, molluscum contagiosum, cysts and warts.

Melanosis vulvae

Melanosis vulvae is a benign condition in which there is patchy, multifocal, dark brown hyperpigmentation of the mucosal surface of the introitus (Figure 1). It can be post-inflammatory

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or spontaneous. Its significance is that it comes into the differential diagnosis of melanoma, and should be biopsied to rule out this possibility.

Melanoma and pigmented naevi

Malignant melanoma presents as an irregular, raised or macular hyperpigmented lesion (Figure 2). Unlike melanosis vulvae, it is not multifocal, but may be poorly defined and very subtle.

Pigmented naevi of the vulva often have an alarming appearance mimicking melanoma (Figure 3). For this reason, any new hyperpigmented lesion on the vulva should be biopsied.

SCC and BCC

Both squamous cell carcinoma and basal cell carcinoma rarely affect the vulva. They present, as on the rest of the skin, with an indurated nodule that may ulcerate (Figure 4). Any such lesion should be biopsied.

Hidradenoma papilliferum is a benign tumour that may present on the vulva with exactly the same appearance (Figure 5).

Seborrhoeic keratoses

Seborrhoeic keratoses are benign lesions that are more common with advancing age. They are common on the vulva as on the rest of the skin (Figure 6). Seborrhoeic keratoses may present diagnostic confusion with warts and squamous cell carcinoma. If there is doubt (e.g. when the lesion is indurated) a biopsy is indicated.

Angiokeratoma

Angiokeratomas are very common, small angiomas found on the labia minora. They present as multiple, small purple papules (Figure 7). They can usually be diagnosed clinically and no treatment is necessary.

Molluscum contagiosum

Molluscum contagiosum is a common viral condition that may be transmitted sexually in adults. The appearance of a papule with a central core is typical (Figure 8). Lesions may be removed by curettage or cryotherapy.

Cysts

Vulval cysts are mainly due to blocked pilosebaceous glands in the hair-bearing areas or sebaceous cysts (Figure 9). Inclusion cysts, usually following obstetric repairs, are less common.

Patients who request excision of uncomplicated cysts should, in our opinion, be counselled against this as surgical excision can be complicated by infected postoperative sinuses.

Sebaceous cysts are asymptomatic except when secondarily infected or when irritated or squeezed. When a patient presents with chronically infected sebaceous cysts, we instigate a regimen of antibacterial soap, low dose tetracyclines, and avoidance of squeezing. Regression in these cases is the rule but often takes many months.

Warts

At present, vulval warts are the most common sexually transmitted disease in Australia. The warts themselves are invariably asymptomatic and are usually discovered accidentally by the

Table. Causes of an asymptomatic vulval rash or lesion

VIN III (vulval intraepithelial neoplasia)	Basal cell carcinoma
Lichen sclerosus	Seborrhoeic keratoses
Melanosis vulvae	Angiokeratoma
Malignant melanoma	Molluscum contagiosum
Pigmented naevi	Cysts, including sebaceous
Squamous cell carcinoma	Warts

IN SUMMARY

- **Causes of a vulval rash or lesion in the absence of other symptoms include VIN III, lichen sclerosus, melanosis vulvae, benign and malignant tumours, molluscum contagiosum, cysts and warts.**
- **Any lesion suspected of being a melanoma or squamous or basal cell carcinoma should be biopsied.**
- **A patient with vulvitis but without macroscopic warts should not be assumed to have symptoms attributable to the human papilloma virus – this concept has been disproven.**
- **Vulval pain, burning and/or unexplained dyspareunia may be due to atrophic vaginitis, referred pain, neuropathic pain, psychogenic pain, vestibular hypersensitivity and/or vulval varices.**
- **Symptoms due to vaginal atophy should respond to topical oestrogen therapy within one month; if not, seek another cause.**
- **Vestibular hypersensitivity does not respond to medical management; biofeedback using pelvic floor exercises is the treatment of choice.**

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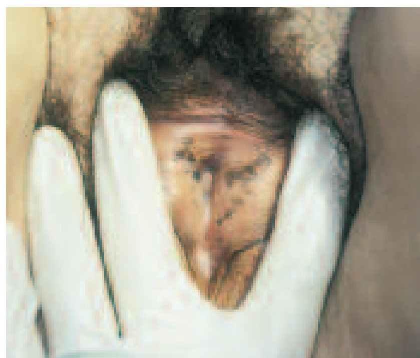

Figure 1. Melanosis vulvae. Multifocal hyperpigmented patches. A biopsy is required to rule out malignant melanoma.

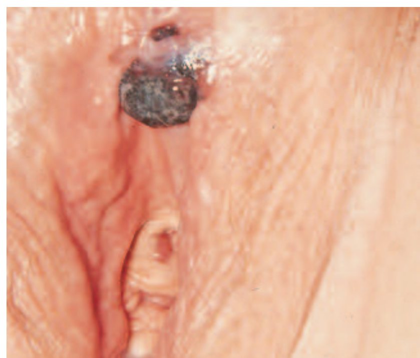

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Figure 2. Malignant melanoma.

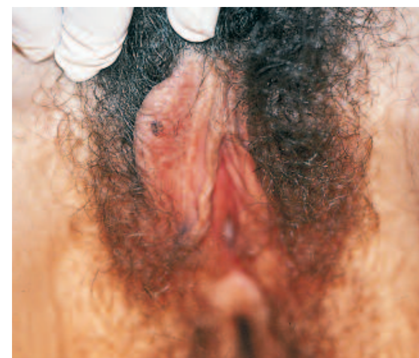

Figure 3. Pigmented naevus of the labium majus.

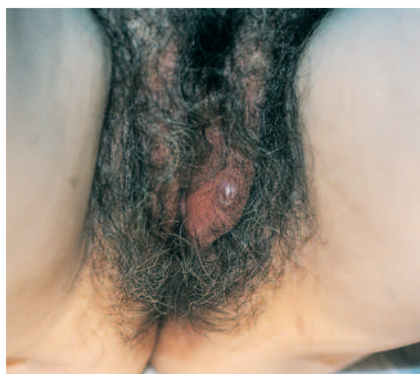

Figure 4. Vulval tumour that proved to be a basal cell carcinoma. Lesions like this should be biopsied.

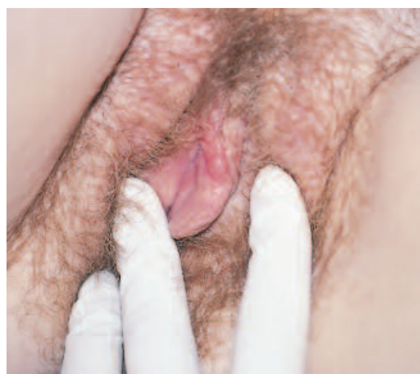

Figure 5. Hidradenoma papilliferum.

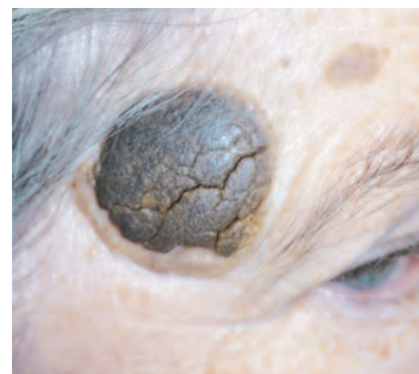

Figure 6. Seborrhoeic keratosis.

patient or doctor (Figure 10). Any associated skin symptoms should be assumed to be due to other causes.

Most patients request treatment for aesthetic reasons and therefore it is imperative that the patient understands that recurrence cannot be prevented by ablation of macroscopic disease. Many crops of warts will regress spontaneously over a period of several months and it is worth trying to convince a woman to wait for three months especially if treatment will require a general anaesthetic.

Patients must have a Pap smear but can, in general, be reassured that vulval warts are a benign condition.

Transmission of the causative human papilloma virus (HPV) is an important counselling issue. Condoms will reduce but certainly not eliminate transmission and in any case the virus is thought to be extremely widespread in the community. There seems therefore no reason to strongly advise the wearing of condoms for the sole purpose of preventing transmission of the HPV. It must be borne in mind, however, that screening for other sexually transmitted diseases should be offered where appropriate.

Warts may be treated with cryotherapy, podophyllotoxin (Condyline Paint) or the new imiquimod (Aldara) cream.

'HPV vulvitis'

During the 1980s it was common for patients with chronic vulval symptoms to be diagnosed as having 'HPV vulvitis'. An assumption was made that 'subclinical HPV infection' was responsible for symptoms, since these patients did not actually have macroscopic warts. The diagnosis was made on the basis of acetowhitening of the vulva and on vulval biopsy. The flaw in this practice was that aceto-whitening is a nonspecific sign on the vulva and the criteria used to diagnose HPV on vulval biopsy were very slim compared with the criteria used on skin elsewhere.

Not surprisingly, treatment aimed at eradicating warts in these patients, most of whom simply had dermatitis, was at best unsuccessful and at worst complicated by exacerbation of symptoms. A

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patient with vulvitis but with no macroscopic warts, even where there is a prior history of them, should never be assumed to have symptoms attributable to HPV. The concept has been disproven and should be abandoned.

HPV and cancer

The true relevance of the finding of vulval HPV relates to its possible role as a carcinogen, particularly in smokers and immunosuppressed patients. HPV has been linked to carcinoma of the cervix and of the vulva and causes condylomata of the cervix, vagina and vulva. Carcinoma of the vulva is uncommon and the link with HPV is not as strong as with the cervix.

The patient with symptoms but no (significant) rash

These patients almost invariably suffer pain and burning, or unexplained dyspareunia.

Patients who complain of pain should be carefully examined to make sure there is no subtle evidence of skin splits, fissures or erosions which, even if minor in appearance, can be very painful. Oestrogen deficiency should be considered. They should always have a vaginal swab to rule out candidiasis, and a trial of therapy aimed at a diagnosis of subclinical dermatitis should be undertaken. If a rash is present this should be treated according to the diagnosis.

If therapy is unsuccessful in eradicating the pain, despite resolution of the rash, and swabs are negative, the patient may be suffering from one or more of the conditions described below. The flowchart on page 46 presents a guide to the differential diagnosis of chronic vulval conditions that present with symptoms but no (significant) rash.

Atrophic vaginitis

It has long been appreciated that vaginal atrophy may cause symptoms such as:

- superficial dyspareunia
- minor vaginal bleeding
- pain from splitting due to friction.

The vaginal mucosa is pale and appears dry (Figure 11). This condition occurs not only in postmenopausal women but also postpartum and in women who are breastfeeding.

Symptoms due to vaginal atrophy alone will improve, promptly and reliably, after one month of topical oestrogen therapy, and vulval or vaginal symptoms that persist after this time should be assumed to be due to other causes.

Newer forms of topical vaginal oestrogen deliver a therapeutic dose in much smaller volumes than older products, and the oestrogen should be one that is poorly absorbed systemically (that is, oestriol [Ovestin cream, Ovestin Ovula pessaries]).

Referred pain

From the back

Not uncommonly, we see patients in whom physical examination is normal but who complain of the sudden onset of vulval pain, either unilateral or more severe on one side than the other side. Usually, by the time we see the patient their history has been complicated by numerous unsuccessful treatments.

A detailed history of a typical patient reveals that the pain was often of sudden onset, related to physical activity. (One patient described lunging to answer the phone.) Thereafter the pain, which has a burning quality, was exacerbated by physical activity and sitting, and relieved by rest and lying down. Often there is a past history of back injury, or an ongoing history of low back pain.

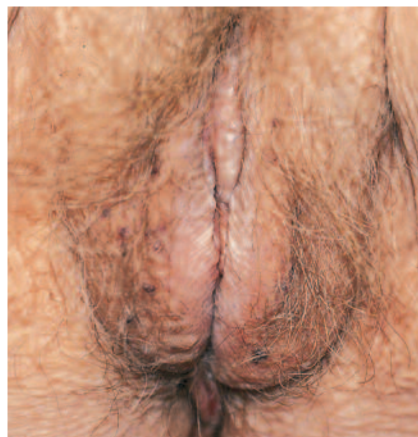

Figure 7. Vulval angiokeratomas.

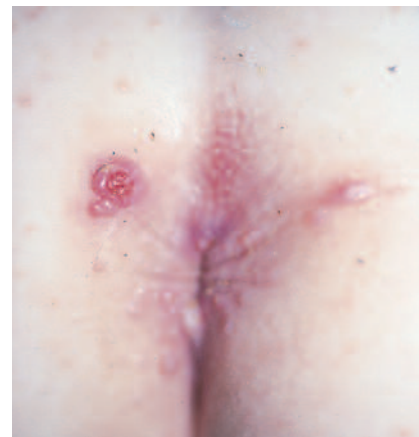

Figure 8. Molluscum contagiosum.

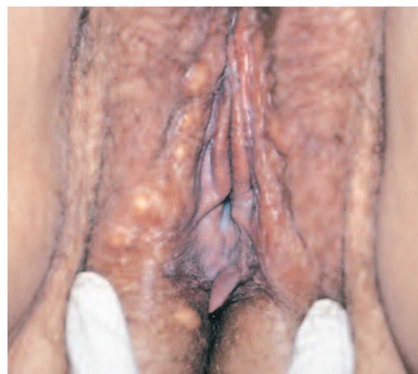

Figure 9. Sebaceous cyst of the vulva.

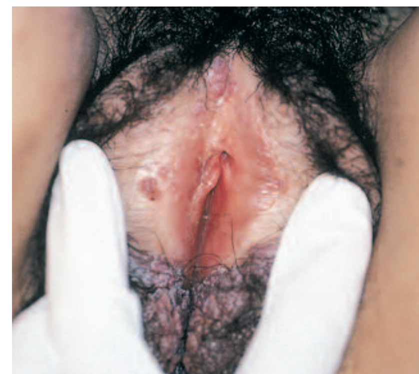

Figure 10. Genital warts.

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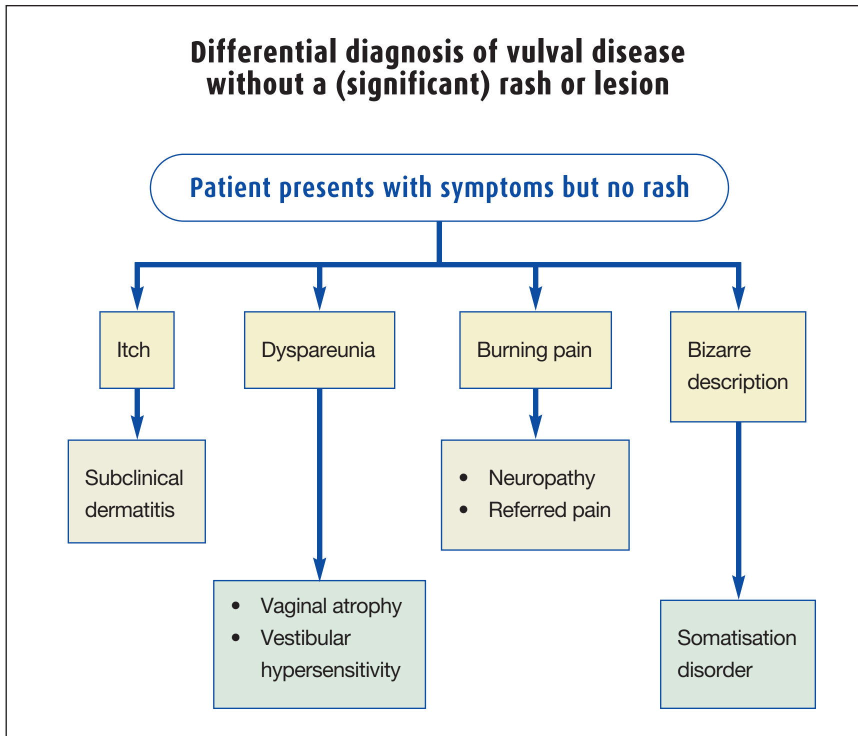

These patients have referred lower back pain, which can usually be relieved by appropriately treating the primary problem. In our experience, chiropractic therapy is often successful.

From the pelvis – gynaecological, urological and colonic pain

Pain localisation below the pelvic floor tends to be poor and the pelvic structures are in very close proximity to one another. Therefore, there are many potential causes of referred vulval pain.

Uterovaginal prolapse often causes dragging pelvic pain that can be perceived by the patient as vaginal pain. Specific posterior wall prolapses always cause varying degrees of vaginal or rectal discomfort. Diagnostic confusion can be cleared by inserting a suitable vaginal ring pessary for one or two weeks to see if reduction of any prolapse relieves the symptoms.

Adnexal gynaecological pathology does not usually give rise to referred vaginal or vulval pain but may do so in conditions which cause inflammation and/or adhesions to the pouch of Douglas – for example, uterosacral endometriosis.

Any anorectal disorder may lead to vaginal pain and this may be perceived as vulval discomfort.

Patients presenting with vulval symptoms also frequently complain of urinary frequency of dysuria and vice versa. The precise aetiological link between bladder and vulvovaginal symptoms is still not clear; however, it seems reasonable to postulate that pelvic floor muscle spasm originating from one part of the pelvis may stimulate dysfunction in another part. We have often observed how severe vulvovaginits can temporarily exacerbate chronic bladder instability.

Patients with deep poorly localised pain, deep dyspareunia and associated bladder symptoms should be referred for gynaecological assessment.

Neuropathic pain

Neuropathic pain, commonly known as ‘dysaesthetic vulvodynia’, is seen most often in older women. The aetiology of this condition is unknown, and is best explained as a neuropathy of pelvic sens - ory nerves of unknown cause.

Symptoms and signs

Patients present with a normal looking vulva for their age but with a complaint of unremitting, poorly localised vulval burning. Sometimes they also experience dysaesthetic sensations such as tingling and formication. The sensation is bilateral and not uncommonly radiates to the inner thighs. There may be a history of back injury.

Although dyspareunia is not a feature, the condition often gives patients a distaste for an active sex life. Sensations

Figure 11. Atrophic vaginitis.

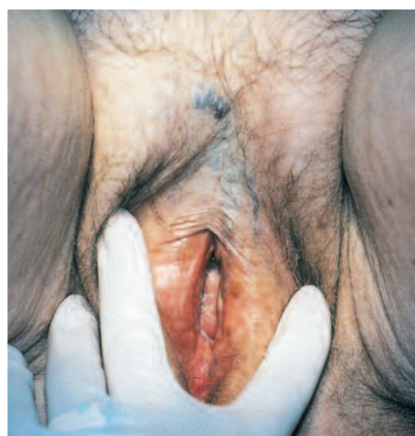

Figure 12. Vulval varices.

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continued

are usually least severe in the morning, intensifying during the day with increasing activity, and settling with rest, similar to sciatica. A similar condition called ‘pudendal neuralgia’ has been described in younger women, in whom dyspareunia and hyperaesthesia feature.

Management

There is often a good response to a low dose (20 to 50 mg) of amitriptyline (Endep, Tryptanol) taken at night. We have also found spinal manipulation helpful in some patients.

It is most important to recognise that even though there is nothing to see, these patients have a real and distressing problem. Often they will have been dismissed as having psychological problems in the past, and for this reason when prescribing a tricyclic it is essential to explain that the tricyclic is being used as a pain relieving, rather than as a psychotropic, drug.

Psychogenic pain (somatisation disorder)

In our experience, psychogenic pain is the rarest cause of vulval pain. It is a very difficult diagnosis to make and in many cases has to be one of exclusion.

Symptoms and signs

The patient gives a history that is bizarre, often with inexplicable periods where she is completely free of the problem or else the pain may extend to other distant areas of the body. These patients often present as depressed, and occasionally as delusional. Either the physical examination is normal or the symptoms are disproportionate to physical findings.

Management

Ideally, these patients require psychiatric help but the nature of their disorder is that they have somatised an underlying psychiatric condition and for this reason any suggestion that they see a psychiatrist is usually rejected. In this circumstance, helping these patients may be impossible and the best approach may be instead to help their friends and relatives cope with their ongoing complaint and minimise medical interventions.

Vestibular hypersensitivity

The hallmark of vestibular hypersensitivity is the sole symptom of dyspareunia. Vestibular hypersensitivity is a relatively common condition. Huge amounts have been written on this subject in the medical literature, not to mention the internet – where there are whole web sites devoted to it.

The unfortunate term ‘vulvar vestibulitis syndrome’ is the most common name used to describe this condition. We say unfortunate, because it is not an inflammatory condition and no amount of medical treatment is ever of value.

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Symptoms and signs

It is common for patients with any sort of vulval inflammatory condition to experience a secondary superficial dyspareunia that resolves with treatment of the underlying problem. It is also not uncommon for patients whose underlying vulval condition has long since been treated to have residual dyspareunia.

However, there is a second group of women who have no underlying condition but who experience superficial dyspareunia, localised to the vestibule that may make sexual intercourse impossible. The typical patient, a young woman in her twenties, gives a history of having been sexually active without pain for a varying degree of time. The onset of the dyspareunia is often sudden and, as time goes by, hypersensitivity to any sort of stretch or pressure becomes more severe, culminating in not even being able to insert a tampon or wear tight jeans. These women are otherwise symptom-free. It can be surprisingly difficult to tease out this history as embarrassment often leads patients to 'beat around the bush' for some time until a direct question about sexual difficulties is asked.

These two groups of women have the same condition; however, in the first the trigger was a physical event and in the second the trigger is an unknown, but we believe a psychological, one in many cases.

Management

Sex is presented in popular literature as always enjoyable and never a source of pain. The experience of pain with sex therefore provokes a great deal of anxiety, and bewilderment. Furthermore, since so many genuinely physical problems in women have in the past been incorrectly ascribed to 'nerves', it has become quite politically incorrect to suggest that there could be a psychological component to this particular problem. Thus these patients have usually tried, unsuccessfully, huge numbers of the various other treatments.

A surgical intervention known as vestibulectomy has been used to treat this condition and some studies have claimed an 80% success rate; however, the procedure is rarely performed in Australia. Given the aetiology of the condition, we have not recommended surgical intervention to our patients.

In our experience, explanation, strong positive reassurance, pelvic floor exercises and biofeedback therapy have been the most successful approaches for this condition. These patients develop higher than normal pelvic muscle tone and this spasm (formerly known as vaginismus) appears to be instrumental in causing pain when the vagina is put on stretch. The response is reflexic but often can be 'unlearned'.

Vulval varices

Vulval varices present as dilated veins, similar to varices in the legs (Figure 12). We have no doubt that vulval varices cause pain by exactly the same mechanism and with the same clinical pattern as varicose veins in the legs.

Diagnosis requires a high degree of clinical suspicion and a willingness to examine the patient's vulva in the erect (standing) position.

Treatment is often unsatisfactory. Pressure from a thick pad within tight underwear does not usually give much relief and surgery is not successful because of the rich venous anastomoses in the area. Sclerotherapy may be helpful.

Conclusion

The differential diagnosis of an otherwise asymptomatic rash or lesions includes both benign and malignant conditions; where there is doubt as to the specific diagnosis, a biopsy is indicated.

Chronic vulval pain, burning and dyspareunia in the absence of obvious abnormality are still poorly understood, but are an important cause of morbidity. Patients with this problem deserve understanding, acceptance and rational attempts to treat their condition. MT

Further reading

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