



HYDROA VACCINIFORME

What are the aims of this leaflet?

This leaflet has been written to help you understand more about hydroa vacciniforme. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

What is hydroa vacciniforme?

Hydroa vacciniforme is an extremely rare skin condition in which there is an abnormal sensitivity of the skin to sunlight (*photosensitivity*). It is neither infectious nor dangerous, but it can restrict an affected person's lifestyle, particularly during the summer months and on holidays.

The term *hydroa* is possibly from the Greek for 'watery eggs', a reference to the blisters that characterise this condition; *vacciniforme* derives from the Greek for the 'pox-like' permanent scars (resembling large deep chicken pox scars) that result when the blisters heal.

Hydroa vacciniforme usually affects children aged 3-15 years, and is more common in females than males. In boys, hydroa vacciniforme may develop at a later age than in girls, and go on for longer.

What causes hydroa vacciniforme?

The cause is unknown. The sun-sensitivity is usually to long wavelength ultraviolet radiation (UVA); it is unclear how this causes the skin problems. Rarely, hydroa vacciniforme has been associated with Epstein-Barr virus infection (the virus that normally causes glandular fever).

Hydroa vacciniforme is not contagious and therefore you cannot catch it from an affected person. There is no evidence that it increases an individual's susceptibility to skin cancer.

Is hydroa vacciniforme hereditary?

Hydroa vacciniforme does not appear to be inherited, but there have been very rare reports of a number of family members being affected.

What are the symptoms of hydroa vacciniforme?

After a short exposure to sunlight (usually between 30 minutes and 2 hours) a tingling discomfort (burning, itching or stinging sensations) develops in the skin, followed by the appearance of lumps (papules) and blisters ('hydroa'). This mainly involves sun-exposed sites, particularly the face, ears and the backs of the hands, although covered sites may sometimes be affected. Occasionally, individuals with hydroa vacciniforme may also experience mild irritation of the eyes, an aversion to sunlight (*photophobia*), a feeling of being generally unwell, and lifting of fingernails and toenails (*onycholysis*).

These symptoms can occur throughout the year, but they are usually worse during the spring and summer months.

What does hydroa vacciniforme look like?

The papules and blisters are of varying size, and the surrounding skin is usually red and inflamed. Over a period of days, they become scabbed and crusted and eventually heal to leave permanent pale depressed scars.

How is hydroa vacciniforme diagnosed?

Hydroa vacciniforme can often be diagnosed from the patient's (or parent's) description and by examination of the skin (or photographs of the rash). If there is uncertainty about the diagnosis, your dermatologist may suggest blood tests and perhaps special tests (*photo testing*). Photo testing is conducted with specialised equipment, and involves different doses of ultraviolet and visible light being shone onto the back of the person being tested to see how sensitive the skin is to light.

Can hydroa vacciniforme be cured?

No, but it is a condition that tends to improve in late adolescence and early adulthood, and usually disappears spontaneously, although the scars are permanent.

How can the rash of hydroa vacciniforme be prevented?

- **Sun-protection.** Many affected people find that protecting their skin from the sunlight by wearing suitable clothing, a hat that protects your face, neck and ears, using a broad-spectrum sunscreen and avoiding sunlight between 10am and 3pm can help prevent the rash. It is best to use a sunscreen with both a high protection SPF (SPF 30 or more), to protect against UVB, and a UVA star rating of at least 4 stars (or look for the circle logo). This should be applied generously (15 to 30 minutes before going out in the sun) and then reapplied every two hours, as well as straight after swimming and towel-drying, during sunlight exposure. Your doctors will advise you about appropriate sunscreens. Most patients can judge how long they can stay in the sun before needing to cover up or seek the shade.
- **Desensitisation.** Some patients may find a desensitisation course of artificial light (narrowband UVB phototherapy) helpful to 'toughen-up' the skin and reduce the likelihood of blisters. This usually involves treatment with light (phototherapy) two or three times a week for about five weeks early in the year (end of Winter). This may help reduce the sun sensitivity which is usually worse in the Summer months, and may also allow individuals with hydroa vacciniforme to stay out longer in the sunlight during the Summer months. The effect is lost over the Winter months and so desensitisation has to be repeated yearly. Occasionally, treatment with a stronger form of light treatment (PUVA phototherapy) may be suggested (see Patient Information Leaflet on [phototherapy](#)).
- **Oral treatment.** Sometimes tablet treatments, used together with sun-avoidance measures, may be helpful. The most commonly used drugs include antimalarial agents (such as [hydroxychloroquine](#)) and beta-carotene (a naturally-occurring substance found in vegetables and fruit). Medicines that dampen down the immune system (immunosuppressants), such as [azathioprine](#) and [ciclosporin](#), may also be considered in some cases. Some studies have suggested that fish-oil supplements may be helpful.

How can hydroa vacciniforme be treated?

If blisters develop, the affected person should cover up and stay out of the sun. The application of steroid ointments or creams, as recommended by your doctor, can help to reduce the redness and discomfort. Additionally, the use of moisturisers will soothe the skin.

Self care (What can I do?)

- Good sun protection as outlined above is most important.
- When you go on holiday, do not forget to take any treatments that have been recommended or prescribed for you or your child.
- If strict sun protection is undertaken, Vitamin D supplements may be required.

Vitamin D advice

The evidence relating to the health effects of serum Vitamin D levels, sunlight exposure and Vitamin D intake remains inconclusive. Avoiding all sunlight exposure if you suffer from light sensitivity, or to reduce the risk of melanoma and other skin cancers, may be associated with Vitamin D deficiency.

Individuals avoiding all sun exposure should consider having their serum Vitamin D measured. If levels are deficient (less than 25 nmol/L) or reduced (less than 50-75 nmol/L) they may wish to consider taking supplementary vitamin D3, 10-25 micrograms per day, and increasing their intake of foods high in Vitamin D such as oily fish, eggs, meat, fortified margarines and cereals. Vitamin D3 supplements are widely available from health food shops.

Where can I get more information about hydroa vacciniforme?

Web links to detailed leaflets:

<http://dermnetnz.org/reactions/hydroa-vacciniforme.html>

Link to patient support group:

Support Group for Sun Sensitive People

Web: <http://sun1.awardspace.com>

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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