



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 a 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 'resembling vaccinia (cowpox)' referring to the scars similar to cowpox 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).

Is hydroa vacciniforme hereditary?

Hydroa vacciniforme does not appear to be inherited, but there have been very rare reports of siblings 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, followed by the appearance of lumps (papules) on the skin 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 scars (“vacciniforme”).

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 simply protecting their skin from the sunlight, by wearing suitable clothing, using a broad-spectrum sunscreen and avoiding sunlight between 11am and 3pm, can help prevent the rash. We suggest the use of a sun-block with both a high (at least 30) sun-protection-factor (SPF) and UVA star rating of at least 4 stars. This should be applied generously and frequently 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 two or three times a week for about five weeks early in the year, and this may 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 that is primarily responsible for the orange pigment in carrots and orange fruits). 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, stay out of the sun and wait for the rash to clear. 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.

What can I do?

- Observe basic sun-protection measures, including the avoidance of sun exposure in the middle of the day, the use of adequate sunscreens, and the wearing of sensible clothing, including a wide-brimmed hat. This is particularly important if you travel to sunny climates. Educate affected children about these measures.
- When you go on holiday, do not forget to take any treatments that have been recommended or prescribed for you or your child.

Where can I get more information about hydroa vacciniforme?

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

Support Group for Sun Sensitive People: <http://sun1.awardspace.com>

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: its contents, however, may occasionally differ from the advice given to you by your doctor.

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