



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 tense, swollen 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 lumps and blisters are of varying size, and the surrounding skin is usually red and swollen. 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/skin biopsy).

Photo testing is performed using 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.

A skin biopsy may be taken to exclude other serious conditions characterized by light-induced reactions. This is a procedure involving taking a small skin sample under local anaesthesia. Stitches may be needed to close the wound. A dressing or adhesive bandage is then placed over the site to protect the wound and prevent bleeding.

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?

Prevention is the key factor in hydroa vacciniforme management. It includes following sun safety tips:

- Avoiding sunlight. Spend time in the shade between 11am and 3pm when it's sunny. Seek shade from the sun before the skin has a chance to redden or burn.
- Wearing protective clothing (thick fabric or specific sun-protective clothing, long sleeves, long trousers, wide-brimmed hat) and UV protective sunglasses.
- Apply a broad spectrum sunscreen (SPF 30 or above and 4 or 5 stars on UVA rating). Apply plenty of sunscreen 15 to 30 minutes before going out in the sun, reapply every two hours and straight after swimming and towel-drying. Most patients can judge how long they can stay in the sun before needing to cover up or seek the shade.

Sunscreens should not be used as an alternative to clothing and shade, rather they offer additional protection. No sunscreen will provide 100% protection.

If very troublesome, desensitisation treatment may be considered. Desensitisation (phototherapy) is a way of helping your skin to be less sensitive to light by exposing it to small doses of UVA or UVB light which are gradually increased. The treatment is done in a hospital outpatient department, in a special phototherapy cubicle. It involves multiple visits for about 5 to 8 weeks in the late winter/ early spring so that the skin is ready to cope with the summer sun.

Occasionally, treatment with a stronger form of light treatment (PUVA phototherapy) may be suggested (see Patient Information Leaflet on [phototherapy](#)).

A few people with extensive lesions may still have problems despite the measures listed above. Some tablets 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 the rash has already developed you should:

- Cover up/ stay in the shade as much as you can and as soon as possible.
- Apply steroid creams/ointments, as recommended by your doctor, to reduce the redness and discomfort. Stop applying steroid cream when the rash has gone.
- Use cold compresses – apply a towel dampened with cool tap water or an ice pack to the affected skin. Ice cubes should be placed in plastic bags then wrapped in a towel before applying to the skin.
- Leave the blisters alone – de-roofing them increases the risk of infection and delays healing. If needed, you can cover them with gauze.
- Take pain relief – available over-the-counter if burning/pain is a problem.

Self care (What can I do?)

- Good sun protection as outlined above is most important. Strict avoidance of sunlight can be devastating for your child, therefore understanding the importance of it and how to apply the sunscreen creams correctly is crucial in preventing of new lesions from appearing and having good quality time together.
- 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:

<https://www.dermnetnz.org/topics/hydroa-vacciniforme/>

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

BRITISH ASSOCIATION OF DERMATOLOGISTS

PATIENT INFORMATION LEAFLET

PRODUCED SEPTEMBER 2009

UPDATED JANUARY 2013, FEBRUARY 2016, SEPTEMBER 2019

REVIEW DATE SEPTEMBER 2022