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Epidermodysplasia verruciformis

What is epidermodysplasia verruciformis?

Epidermodysplasia verruciformis (EV) is a rare, autosomal recessive inherited skin disorder characterised by eruptions of wart-like lesions that may occur anywhere on the body. These lesions are caused by infection with the [human papillomavirus](#) (HPV). There are over 70 subtypes of HPV that are responsible for causing a wide range of viral warts. In the general population infection with some subtypes of HPV have minimal or no clinical effect, however in patients with EV, infection with these same subtypes can cause flat, wart-like lesions. It has been found that patients with EV have an abnormal or impaired immune response to HPV or wart viruses. In patients with EV more than 30 subtypes of HPV have been implicated.

A major concern for patients with EV is that the wart-like lesions may transform into [skin cancers](#). The exact reason how these lesions become cancerous is still unclear but exposure to sunlight or UV rays appears to be involved in the progression from benign warts or verrucae to malignant skin cancers. It has also been discovered that the potential for EV lesions to turn into cancers may depend on the type of HPV infection present. Over 90% of EV-associated skin cancers contain HPV types 5, 8, 10, and 47. Meanwhile EV lesions caused by HPV 14, 20, 21, and 25 usually remain benign (non-cancerous) skin lesions.

Who gets epidermodysplasia verruciformis?

EV is an autosomal recessive inherited disorder, which means that 2 abnormal EV genes (one from each parent) are needed to have the disease. It has been found that 10% of patients with EV come from marriages between blood relatives (i.e. the parents share a common ancestor). In a few cases sex-linked and autosomal dominant inheritance patterns have also been found.

The warts usually start to develop sometime during childhood. About 7.5% of cases appear in infancy, 61.5% in children aged 5–11 years and 22.5% in puberty. The disease affects both males and females and people of all races.

What are the clinical features of epidermodysplasia verruciformis?

Two types of EV lesions have been described.

- Flat, wart-like lesion that look like plane warts. These can be flat-topped papules ranging from light pink to violet in colour. In some places papules may join together to form large plaques. These may be reddish brown and have scaly surfaces and irregular borders.
- Verrucous or [seborrhoeic keratoses](#)-like lesions. These are most commonly seen on sun-exposed skin and are often slightly raised, brown coloured lesions.

Flat wart-like lesions also commonly appear on sun-exposed areas such as the hands, feet, face, and ear lobes. Plaque-like lesions usually appear on the trunk, neck, arms and legs. The palms, soles, armpits and external genitals may also be involved.

In most cases the warts occur in clusters and can be anything from a few warts to over a hundred warts.

What is the treatment for epidermodysplasia verruciformis?

EV is a lifelong disease. Even though lesions can be treated or removed as they appear, patients with EV will continue to develop these lesions throughout life. In many cases lesions may develop and remain unchanged for years. The greatest risk is that in 30–60% of EV patients these lesions will change into skin cancers. Malignant tumours are usually found in patients between 30–50 years of age.

Currently there is no treatment to prevent new EV lesions from occurring. The management of EV involves a combination of medical and surgical treatments alongside patient counselling and education.

- Stress the importance of following [sun protection](#) strategies. Exposure to sunlight (UVB and UVA) has been shown to increase the rate of EV lesions turning into skin cancers.
- Treat and remove warts, as you would treat other [viral warts](#). This includes chemical treatment, [cryotherapy](#) with liquid nitrogen and [electrosurgery](#).
- Oral and topical treatment with retinoids ([isotretinoin](#) and [acitretin](#)), [fluorouracil](#) and [imiquimod](#) are proving to be useful agents.
- Experimental therapies for EV lesions and skin tumours include intralesional interferons and retinoids, combination of isotretinoin and interferon alpha or cholecalciferol ([vitamin D](#)) analogues.

Related information

References:

- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.

On DermNet NZ:

- Viral warts
- Seborrhoeic keratoses
- Skin cancer

Other websites:

- [Epidermodysplasia Verruciformis](#) – e-medicine dermatology, the online textbook

Books about skin diseases:

See the [DermNet NZ bookstore](#)

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DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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