

How Ectodermal Dysplasias Can Affect The Skin

Several types of skin-related problems typically occur in ectodermal dysplasias. It is important to understand that the skin issues vary according to the type of ectodermal dysplasia. Following is a description of those that are most common.



Appearance at Birth

At birth, how the skin looks differs depending on the type of ectodermal dysplasia. The skin of infants may be

- dry
- cracked
- pale and typically appears thin in some types of ectodermal dysplasias. The surface blood vessels may be prominent.
- peeling. In infants affected by hypohidrotic ectodermal dysplasia (HED), the skin of the hands, feet, and, sometimes the entire body surface, may have more than usual redness and peel, revealing normal-looking skin underneath. This clue can lead to early diagnosis of ectodermal dysplasia. These changes are similar to those that occur in babies born one to two weeks after their due dates (“post-mature”). There is no simple way to tell these conditions apart at birth. If an infant is not post-mature, and especially if a relative has been identified as having HED, such skin changes suggest the possibility of ectodermal dysplasia. Consult with a pediatric dermatologist or medical geneticist to help establish a diagnosis.
- lax around the eyes
- thick on the palms and soles



Infancy/Toddlers

Diaper Rash

Diaper rash is a common problem for children in diapers. Most often, this condition is caused by irritation from urine and stools, moisture, and overgrowth of germs (bacteria or yeast).

Seborrhea

Many young children have scaly scalp, most often called “cradle cap” or seborrhea. This condition has a variety of causes, including a reaction to a type of normal skin yeast.

Any Age

Skin Coloring

The skin may be pale and typically appears thin in some types of ectodermal dysplasias. The skin may be darkened around the eyes or on the elbows, palms, and soles. With time, the skin of children with HED which is characterized by decreased or absent sweat glands may be shiny, dry and appear thin, with visible bluish underlying blood vessels.

Dry Skin

The skin may be dry, scaly, and easily irritated. Dryness is caused primarily by genetically defective or absent skin surface proteins or skin gland fats that evolved to help trap moisture on the skin's surface. Oil and sweat glands may be poorly developed or absent. External factors like weather and excessive soaping can make it worse.

Skin Infections and Skin Erosions

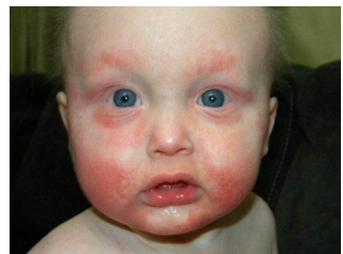
Most people with ectodermal dysplasia do not experience frequent skin infections. However, the skin on the scalp, hands and feet in ankyloblepharon ectodermal dysplasia-cleft lip and/or palate (AEC) syndrome and ectrodactyly-ectodermal dysplasia clefting (EEC) syndrome may become red, weepy, crusty and somewhat swollen.

Eczema

People with HED are at higher risk for a more bothersome form of red, scaly and itchy dry skin called eczema. Eczema is common in people without ectodermal dysplasia, too, and its treatment is much the same for everyone. Eczema may cause a burning sensation or itch that interferes with sleep. Certain areas of the body are more likely to have eczema: the neck, face, hands, feet, crook of the elbows and backs of the knees.

Enlarged Facial Oil Glands

Another skin problem sometimes occurs in males with hypohidrotic ectodermal dysplasia (HED). As they get older, small, flesh-colored bumps on the face often develop, usually around the eyes and on the upper cheeks. These are enlarged oil ("sebaceous") glands.





Sun Safety

Individuals affected by ectodermal dysplasia may be prone to sunburn but can tolerate outdoor activities with routine use of commonly available sun protection products.

Small or Absent Nipples

In both males and females affected by HED, nipples may be small or absent. Moreover, females who are either affected by or are carriers of ectodermal dysplasia may have underdeveloped breasts or breasts that are very unequal in size.

Visit our Library at www.NFED.org to find more detailed information about skin issues in the ectodermal dysplasias and their treatment.