

## Nailing Down

### Some Common Nail Problems in Ectodermal Dysplasias By Alanna F. Bree, M.D.

#### Reproduced from the Educator with the kind permission of the NFED

Just as there are many different types of Ectodermal Dysplasias, there are also many different nail changes that can occur in association with these conditions. The nail is a complex structure that is formed from the layer of the developing body known as the ectoderm. We know that the hair, nails, sweat glands and teeth are formed from the ectoderm, and that is why many people with Ectodermal Dysplasias can be affected by changes in the appearance of their nails.

The nails typically develop in a baby during the 15th week of pregnancy, and the nails, under normal circumstances, grow without stopping until the end of life. The average growth of the fingernail is 3 mm per month and the average growth of the toenail is 1 mm per month.

The fingernails and toenails are produced by actively growing cells in the nail matrix. The nail matrix is under the skin at the base of the nail. If you push down gently on the area of skin just below the bottom part of the nail, you can feel the nail matrix depress slightly with a subtly squishy feel. The nail matrix extends up under the nail and you can see it as the white half moon area, known as the lunula, at the base of the nail.

The nails grow out of the matrix and harden in the shape of a rainbow. The cells that produce the top surface of the nail are the ones that are further back under the skin, and the ones that are within the lunula produce the under surface of the nails. The cuticle is there to protect the nail matrix and to help the nail grow out evenly and smoothly. That is why you do not want to aggressively remove or damage the cuticle because it helps your nail remain healthy and grow normally.

The shape and appearance of the fingernails and toenails can be affected by many different factors. Many people without Ectodermal Dysplasias have abnormal changes to their nails that are caused by a variety of different things which can include medical conditions, illnesses, viruses, injury, repeated trauma, nutritional deficiencies, medications and environmental factors. There are many genetic conditions, including Ectodermal Dysplasias, which can affect the growth and appearance of the fingernails and toenails. Not all people who are affected by Ectodermal Dysplasias will have changes to their nails. In fact, the nails in Ectodermal Dysplasia may be quite normal.



**Nails in XLHED are typically paper thin**

#### **Nail Issues in Ectodermal Dysplasias**

The most common nail changes seen in people affected by Hypohidrotic Ectodermal Dysplasia (HED) are typically thin nails. In Clouston syndrome, the nails can be thick and discoloured with very slow growth, and they can also lift up off the finger. The nail changes in Goltz syndrome are quite varied and can include absent nails, but the most common features are typically ridging and splitting of the nails, along with V-shaped notches at the end of the nails.

The nail changes in AEC syndrome are even more widely variable and include absent or disappearing nails, thinned or thickened nails, irregularly shaped nails, and nails with an uneven edge. EEC syndrome can also affect the nails, which may include absent nails, thickening of the nails or small pits in the nails.



**Nails in Clouston syndrome can be thick and discoloured with very slow growth**

### Treatments

Treating nail problems, even in people without Ectodermal Dysplasias, can be difficult because there are very limited therapies available. For nails that are absent or irregularly shaped, the only potential treatment, if desired, is the application of artificial nails. Artificial nails work well for some people, who are often very pleased with their appearance.

Aside from the cost of the products, and that of professional application if used, is the potential for irritation of the surrounding skin, allergic reactions and further damage to the nail. These complications are most commonly related to the chemicals that are used to adhere the artificial nail to the skin. If these problems develop, the use of artificial nails should be discontinued and they will often improve with time.

### Thin Nails

Nails that are thin or brittle are best treated with the application of an ointment-based moisturizer to the cuticle area and the entire nail on a daily basis. It is also best to avoid extensive exposure to water or harsh chemicals as this can make the nails more brittle. To add a layer of protection to the nail, application of a clear nail polish or lacquer may be helpful. A diet that is very low in protein can also lead to thinning of the nails, so you should have an adequate amount of protein in your diet. There are also some studies, although significant data is lacking, showing that biotin at 2.5- 5.0mg/day may be helpful.



**Nail changes in Goltz syndrome vary but can include absent nails and ridging and splitting of the nails**

### Thick Nails

For thick nails, the nail can be filed on a regular basis and this is best done after soaking in warm water for a few minutes and should be followed by application of a moisturizer. Application of creams with urea or salicylic acid to the nail can also help soften and thin them, but needs to be done on a regular basis to have an effect.

Since these creams can be quite irritating to the skin, it is best to apply an ointment, like Vaseline, to the skin around the nail first to protect it and then apply the medicated cream over the nail only. To help the medicated cream penetrate in the nail and have the most effect, tape or plastic wrap can be applied over it and kept in place overnight.



**Nails in AEC syndrome can include absent or disappearing nails, thinned or thickened nails or irregularly**

### Curved Nails

For nails that are curved and grow into the skin, causing irritation and ingrown nails, it is best to keep the nail and surrounding skin soft with application of a moisturizer daily. The nails should be trimmed straight across, not rounding the edges, and the nails should be kept slightly longer than the end of the fingers or toes, if this is possible. Wearing shoes, as well as tights, pantyhose or socks, that are too tight can worsen this problem and cause further irritation.

You can also gently apply dental floss or small wisps of cotton from a Q-tip or cotton ball under the edge of the nail that will help lift it up slightly and reduce the irritation and discomfort. If this is a significant problem, nail surgery with a dermatologist or podiatrist may be necessary. They can remove a portion of the nail or the entire nail if necessary. If this becomes a recurrent problem, they can apply a chemical to the nail matrix after the nail is removed so that it does not grow back.

### Loose Nails

If the nails are loose, it is best to keep them trimmed to the edge of the fingertip or toe so they do not catch on anything. It is also helpful to keep them well moisturized. The loose nail will typically shed painlessly on its own when the new nail has partially grown in underneath the old nail.

If the nails are lifted up, it is important to soak the nails daily in warm water for a brief time to remove debris from under the nails which will help prevent any infections. Use of sharp instruments to clean under the nails should be avoided as this often leads to further damage. Application of liquid band-aid can also be applied under the nail to aid in keeping the nail in place; although, this is rarely associated with irritation to the skin.

### Discoloured Nails

If the nails are discoloured, it can indicate a bacterial or fungal infection, but it is always best for your doctor to take a clipping of the nail so the sample can be sent to the lab for culture. This is the only way to know if an infection is truly the cause of the nail discoloration or changes you see. A yellow-green discoloration may indicate a bacterial infection and can be treated with daily diluted vinegar soaks (1 tablespoon of white vinegar to 1 quart of water). But, it may also need to be treated with an antibiotic medication prescribed by your doctor, especially if the skin around the nail is red or inflamed.



**These toe nails are smaller in size and have a rough texture**

If the nail is white to yellow-brown with debris under the nail, then you may have a fungal nail infection. Over-the-counter topical antifungal medications may help improve the appearance, but definitive treatment requires an oral antifungal medication that is prescribed by your doctor.

There are a myriad of other nail changes that can occur, but the ones that were reviewed are likely the most common problems that you may encounter. Our nails, just like us, come in a variety of shapes and sizes. They are only a small part of what makes us up on the outside, but remember, it is what is on the inside that defines us as a person. So to that end, here is some advice from Will Rogers, "What the country needs is dirtier fingernails and cleaner minds."

## Supporting a normal lifestyle

Ectodermal Dysplasia Society (Registered Charity No. 1089135). Disclaimer: Any views or opinions are made by the author in good faith. No liability whatsoever is accepted by the author or the Ectodermal Dysplasia Society. Recipients should make their own additional enquiries of medical and other relevant authorities before acting on these views. The use of a product name does not constitute a recommendation or endorsement by the author or the Society.

## **Supporting a normal lifestyle**

Ectodermal Dysplasia Society (Registered Charity No. 1089135). Disclaimer: Any views or opinions are made by the author in good faith. No liability whatsoever is accepted by the author or the Ectodermal Dysplasia Society. Recipients should make their own additional enquiries of medical and other relevant authorities before acting on these views. The use of a product name does not constitute a recommendation or endorsement by the author or the Society.