

Neonates and Infants with an Ectodermal Dysplasia Syndrome

What is ED?

Ectodermal Dysplasia is not a single disorder, but a group of closely related disorders known as the Ectodermal Dysplasias. More than 170 different syndromes (types) have been identified. The Ectodermal Dysplasias are genetic disorders affecting the development or function of the teeth, hair, nails and sweat glands. Depending on the particular syndrome that an individual has, Ectodermal Dysplasia can also affect the skin, the lens or retina of the eye, parts of the inner ear, the development of fingers and toes, nerves and other parts of the body.

What is the Cause?

Ectodermal Dysplasias are caused by altered genes. The altered genes may be inherited or normal genes may become defective (mutate) at the time of conception. The chances for parents to have affected children therefore depend on the type of Ectodermal Dysplasia that exists in the family.

It is important to remember that a person cannot choose or modify the genes that he or she has, and that events of pregnancy do not change the genes. Thus, parents who have a child with Ectodermal Dysplasia should not think that they did anything to cause the defective gene and should not blame themselves for its existence. Genetic counselling is available for families, and you may find this helpful.

How can I look after my baby?

You can do all the things any newborn baby needs – such as handling, feeding, and bathing, but you may need to change the way some of these things are done. The following guidance information should not be relied upon exclusively and it is important to seek a Doctor's advice should any problems be encountered.

Temperature Control

If your child has the form of ED known as Hypohidrotic (“less damp”) Ectodermal Dysplasia, one of the main factors is the lack of temperature control. Individuals

with this condition have an inability to sweat due to absent or reduced sweat glands and therefore overheat at any time of year, either from atmospheric temperatures or a developing infection. This can be dangerous particularly to the very young. Several Ectodermal Dysplasia syndromes experience lack of temperature control as a result of hypohidrosis (lack of sweating) or hyperhidrosis (sweating).

However, the signs and symptoms of jaundice, which is the most common condition requiring medical attention in the newborn period, (e.g. lethargy, poor feeding), could potentially be confused with overheating.

In addition, neonates are also particularly susceptible to becoming cold. Any baby who is already vulnerable such as a poorly or premature baby or one with an infection will find it especially hard to metabolise the fat from which baby sustains its energy.

It is therefore important to consult a health professional if you have any concerns. Your baby's temperature can be easily and frequently checked by your midwife during your stay in hospital, and you will soon learn to recognise what is and isn't normal behaviour for your baby.

Once you have taken your baby home, it is important to train yourself to curb a very natural instinct to wrap your infant warmly (this does not mean keep baby cold) and even harder, I think, to ignore Granny and well-meaning friends and relations. Just keep feeling baby, and judge for yourself; as a mother you will have a natural instinct. However, some individuals have found that the ears of infants with an Ectodermal Dysplasia go very red whilst their body remains pale when they are overheating. As your baby gets older the problems often get easier to deal with, although it will possibly never go away.

If baby has been fed, winded and changed, and is still irritable, fractious and crying it may be that their temperature is rising. It is very difficult to judge a tiny baby's temperature and is often solved by a process of elimination; pull back the blanket or take baby to a cooler environment, if the fractiousness is due to overheating baby will calm down.

For older babies, a fan will help tremendously in winter as well as in summer. It must be remembered children with temperature-sensitive Ectodermal Dysplasia suffer from inside temperatures in the winter months due to central heating, just as much as outside temperatures in the summer months.

If you feel baby is unwell it is important to seek the advice of a Doctor as soon as

possible.

Should any infant or child have a temperature due to an infection, the usual Calpol (paracetamol suspension) will reduce the fever, but care should be taken as the internal core temperature may continue to rise after the use of Calpol. Plenty of flannelling down with tepid water (not cold water) and the use of a fan will help reduce the temperature. It is advisable to seek your Doctor's advice before administering any medication.

Travelling by car or public transport (unless air conditioned) should be avoided in hot weather if at all possible, but if absolutely necessary the following precautions are essential: ensure you have a container of water and a flannel for sponging down, a fan if you have one and plenty of cool drinks. It may also be a good idea to make use of sun blinds to shade the passengers in the back of the car. Always try and park in the shade. Cardboard windscreen covers for when your car is stationary are also extremely helpful.

Clothing

Cotton clothing will help keep baby cool and help prevent skin irritation and rashes. You may find it better not to put a vest on baby, but to use cardigans instead so they can be easily put on and taken off. Sleep suits and sleeping bags are best avoided as baby will not be able to kick them off when they get too warm.

Feeding

Newborn babies with Ectodermal Dysplasia may have difficulty feeding by either breast or bottle. This may be partly due to lack of temperature control, breathing difficulties, swallowing difficulties or weak muscle tone. It is not uncommon for infants with Ectodermal Dysplasia to choke, vomit and take a long time, sometimes as long as an hour, to feed. If baby gets too hot he or she may become fractious and cry; the more baby cries the hotter he or she will become; this may lead you to think that baby is still hungry.

Some ways which may help are:-

Breast feeding - it is advisable not to wrap the baby up too much against the mother's body as baby may get too hot and this may then affect sucking and swallowing ability. Skin to skin contact between mother and baby is highly recommended by health professionals not only to promote breastfeeding and bonding, but also because the mother has a unique ability to regulate baby's temperature, breathing and heart rate. It will also pass the mother's skin flora to the baby, protecting against infection.

Female carriers of X-Linked Hypohidrotic Ectodermal Dysplasia can have underdevelopment (hypoplasia) of the nipples which can add to the baby's feeding problems; this may be overcome with a lot of perseverance, but it may be difficult.

Bottle feeding - Feeding little and often can help as does not heating the milk too much; give at room temperature instead. As with breast feeding it is best not to wrap baby up too much and if possible to lay baby on your lap instead so baby doesn't get too hot.

To help prevent choking, the level of milk flow is important, too fast and baby is unable to take enough breaths, too slow and sucking becomes harder and baby tires quickly. Any concerns can be discussed with an infant feeding advisor, who is available to all mothers whether breast or bottle feeding. However, bottles are available where baby decides the flow of milk. Using this type of bottle may prevent some of the issues of choking etc., which are found with the old type of bottles where too much milk is released into baby's mouth.

Breathing/Swallowing – The mucus in the nose and throat may be thick and not drain properly causing baby to continually have blocked nasal passages. It is important to ensure the nasal passages are cleared of mucus regularly and especially before feeding; a suction machine may be used as mentioned below. While feeding, baby sucks, stops breathing to swallow, then starts breathing again. If baby has nasal congestion, breathing will be difficult thus interrupting the pattern of feeding and possibly causing choking.

Crying/Voice –baby's cry may sound different from other babies; this may be due to hoarseness caused by mucus in the larynx (voice box) or dry throat and respiratory tubes.

Vomiting, Projectile Vomiting and Choking – This is fairly common in infants with Ectodermal Dysplasia; there are several possible reasons for this, such as weak throat muscles, struggling to breathe properly due to blocked nasal passages, overheating etc.

Skin

This may be pale and transparent with veins clearly visible and there is often increased pigmentation around the eyes or on the elbows, palms and soles of the feet. The heavily pigmented skin around the eyes may be wrinkled and that on the palms and soles may be thick. At birth the skin of a newborn is often similar to that of a "post-mature" baby with increased redness and peeling. This clue can lead to early diagnosis of Ectodermal Dysplasia. Nappy rashes may be persistent later in infancy.

Very dry skin is without exception a problem. There is a wide range of products available to help prevent and treat dry skin, but it may be best to avoid very greasy products in hot weather. Neither soap nor any perfumed bath additives, including baby bath products, should be used as these all have a drying effect and are irritating to the skin which may be sensitive. Frequent bathing keeps the skin moist. Always pat dry and apply cream immediately. In a large number of cases eczema is a problem and for some it is extreme. This must be treated by your doctor. During the summer months individuals with fair skin should use a high factor sun block at all times, preferably one for sensitive skin.

Scalp crusting, similar to 'cradle cap' may be experienced; it is advisable to seek your Doctor's advice.

Nasal Congestion

This is a common problem and happens because the mucus membrane may be malformed. The mucous secretions are much thicker than normal, forming a crusty mass.

It is extremely helpful if a suction machine can be borrowed from the hospital when baby is discharged as this is the best method for clearing the nasal passages in such a tiny baby and will help with feeding. However, it is extremely important to obtain specific training and instructions to ensure there is no harm caused to the baby's nasal passages.

For small babies the doctor can prescribe Minims which are saline water eye drops, but are packaged in such small quantities that they are excellent for use as nose drops. A few drops a day is all that is needed to help keep baby's nasal passages moist and thereby make breathing and feeding easier. There are as several nasal aspirators on the market which can be purchased from baby stores, chemists, etc.

It is possible to buy or have your doctor prescribe nose drops, but vasoconstrictors e.g. ephedrine should not be used long term. However, the long term use of saline water drops/spray is fine.

Suction units are available via the internet. One product that has been recommended by our members can be found at <http://www.4little1.com/baby-nose-clear/nasal-aspirator-baby-nose-clear>

Respiration

The linings of the nose, larynx, trachea and lungs are moistened by various glands, some of which may be absent, reduced in number or may not function normally.

The underproduction of respiratory fluids together with constant nasal congestion can cause frequent infections for which antibiotics maybe necessary. Croup is a common problem in younger children and asthma occurs frequently in individuals of all ages. Both should be referred to your doctor without delay.

It may be helpful to have a humidifier in the bedroom at night to help moisten the air which will help keep the respiratory tract and nasal passages moist.

Eyes

Most people with Ectodermal Dysplasia have normal vision and appear to have no greater need for glasses than anyone else. However, visual problems in Ectodermal Dysplasia may be caused by lack of tears, infections (conjunctivitis), corneal scars, cataracts and retinal changes. Dry eyes are a common problem and artificial tears may be necessary. Crusting of the eyes can also be an issue and if this happens bathe the eyes gently with tepid sterile water, remembering to wipe once from the inside to the outer corner of the eye and discarding each piece of cotton wool once used. Sensitivity to light (photophobia) all year round can be painful for dry eyes; sunglasses or tinted glasses will help when baby is older, or try to keep baby in the shade. Irritation to the eyes may be caused by eyelashes which grow in an abnormal direction; this can be confirmed by an Optician and treated by an eye specialist.

Teeth

One of the main characteristics of Ectodermal Dysplasia, and the one on which a diagnosis is frequently made or proven, is the absence of some of the teeth (Hypodontia). Ectodermal Dysplasia can affect the primary (first) and permanent teeth, but is more common in the latter. Occasionally all the teeth may be absent (Anodontia), although this is very rare. Teeth that are present are often smaller than normal, widely spaced and often pointed. In some cases, the enamel is also defective and discoloured. The teeth often erupt later than normal in people with Hypodontia, especially if many are absent. Due to this babies with Ectodermal Dysplasia often do not have any teeth showing until they are around two years of age.

Some dental hospitals have specialist teams to help patients with these kinds of dental problems, and can provide a coordinated treatment plan involving all the necessary experts. Ask your dentist about being referred to one, if this facility is available in your area, or seek the help of the Ectodermal Dysplasia Society.

Hair

The scalp hair may be absent, sparse, fine, lightly pigmented (very blond), or

abnormal in texture. The hair may also be fragile and unruly, stick out in all directions, get extremely knotted and difficult to comb. The hair is dry because the oil glands are absent or poorly developed. As many shampoos may dry the hair and scalp it may be better to wash with bath oil which has preferably been prescribed by the Doctor as opposed to a brand shampoo. A medicated shampoo may be too strong for the scalp and be best avoided.

Ears

Hearing loss may occur due to hard dry impacted wax possibly caused by the underproduction of body fluids. It is advisable to seek your Doctor's advice and maybe use Olive oil drops to help soften the wax.

Throat

Saliva is sparse, causing problems with chewing, tasting and swallowing foods. It is advisable to avoid dry foods and always have a drink to hand when eating. A hoarse, raspy voice and cry is common.

Nails

The nails may be poorly developed, small, thick or thin, brittle, discoloured, cracked, or ridged. In addition, they may grow slowly, are shed periodically, and develop light spots, lines or patches. It is also possible for nails to become infected by fungus or yeast and to have a bad odour. Seek your Doctor's advice as treatment may be needed. Generally, nails should be kept short and moisturised daily with something similar to Vaseline.

Other

It should be said that not everything that affects an individual with Ectodermal Dysplasia is as a result of having Ectodermal Dysplasia and that it is possible to have other conditions or symptoms as well. For example, it is possible that some speech problems are attributable to Ectodermal Dysplasia, but a stammer would be a separate problem. It is always advisable to seek your Doctor's advice.

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