

The First Years – Neonates and infants affected by an Ectodermal Dysplasia Syndrome

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 information is for guidance but should not be relied upon exclusively and it is important to seek a Doctor's advice should you encounter any problems.

Temperature Control

One of the main factors is the lack of temperature control in the form of hypohidrosis (lack of sweating) or hyperhidrosis (increased sweating). This can be dangerous particularly for the very young. Please look at our website www.edsociety.co.uk for more information on hypo and hyper - hidrosis.

However, do note that the signs and symptoms of jaundice, a very common condition requiring medical attention in the newborn period, could potentially be confused with overheating as jaundice can also result in lethargy and poor feeding. In addition, neonates are particularly susceptible to becoming cold. It is therefore important to consult a health professional if you have any concerns about your baby. 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 baby warmly (this does not mean letting baby get cold) and it can be even harder to ignore Granny and well-meaning friends and relations. 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 it is often solved by a process of trial and error: pull back the blanket or take baby to a cooler environment. If the fractiousness is due to overheating, baby will calm down. Just keep feeling your baby and judge for yourself. However, some individuals have found that the ears of infants affected by 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 the problem will possibly never go away.

For older babies, a fan will help tremendously in winter as well as in summer. It must be remembered that children affected by 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 your baby is unwell it is important to seek the advice of a Doctor as soon as possible.

Should any infant or child have a high 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. However, if the use of Calpol does not reduce the fever it is advisable to seek your Doctor's advice or that of the A&E hospital department.

Travelling by car or public transport (unless air conditioned) should be avoided in hot weather if 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 affected by 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 affected by Ectodermal Dysplasia to choke, vomit and take a long time, sometimes as long as an hour, to feed. If baby gets too hot, they may become fractious and cry; the more baby cries the hotter they will become; this may lead you to think that baby is still hungry rather than too hot.

Some ways which may help are:

Breast feeding - it is not advisable to wrap the baby up too much or cradle baby too close to mother's body as this may make them too hot and may then affect their ability to suck and swallow. It may be best to place baby on your lap and use a fan.

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. However, it is advisable to do this when not feeding baby.

Females who are carriers of X-Linked Hypohidrotic Ectodermal Dysplasia may have underdevelopment (hypoplasia) of the nipples and breasts, 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 it at room temperature instead (not at body temperature). 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 the 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 feeding by breast or bottle. 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. Choking may also be experienced if baby is too hot.

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 and Choking – This is fairly common in infants affected by 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. However, vomiting that is sustained and forceful could be a cause for concern and it is advisable to seek your Doctor’s advice promptly if this occurs. Repeated bouts of less severe vomiting might also be a problem if baby is not gaining weight properly.

Skin

At birth the skin of a newborn affected by Ectodermal Dysplasia is often similar to that of a “post-mature” ("over-cooked") baby with increased redness and peeling. This clue can lead to early diagnosis of Ectodermal Dysplasia. Nappy rashes may be persistent later in infancy. The skin 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.

Very dry skin is usually 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 the skin and apply cream immediately. In a large number of cases eczema is a problem, and for some it is extreme and must be

treated by your Doctor. During the summer months a high factor sun block should be used at all times, preferably one for sensitive skin.

Scalp crusting, similar to 'cradle cap' may be experienced; this can be mild or severe. 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 that using the machine causes no harm to the baby's nasal passages.

For small babies the Doctor can prescribe Minims, which are saline water eye drops. Because they are packaged in such small quantities, 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 several nasal aspirators on the market that 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) are usually of no benefit (the benefit will usually come from the fluid not the ephedrine) and their long-term use is harmful. However, the long-term use of saline water drops/spray is fine.

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 not functioning normally. The underproduction of respiratory fluids together with constant nasal congestion can cause frequent infections for which antibiotics may be necessary. Croup is a common problem in younger children and asthma occurs frequently in individuals of all ages. Individuals with either an infection or croup should be referred to a Doctor without delay.

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

Teeth

Babies affected by Ectodermal Dysplasia often do not have any teeth erupt until they are around two years of age. Those that erupt may be widely spaced, conical and in some cases the enamel is defective and discoloured. Hypodontia (missing teeth) may also be experienced.

Some dental hospitals have specialist teams to help children with such dental problems and can provide a coordinated treatment plan involving all the necessary experts. Ask your Dentist about being referred to a dental hospital, if this facility is available in your area, or seek the help of the ED Society.

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.