

ED and Ear, Nose and Throat Problems

by

Richard Mills, Senior Lecturer and Consultant ENT Surgeon
University of Wales College of Medicine

In clinic, recently, I had two medical students with me. Knowing that the next patient I was to see with them had Ectodermal Dysplasia, I thought I would say a word or two about the condition and ask them what problems, both in general and in ear, nose, and throat (ENT) terms, they might expect the patient to have. They had not heard of the condition. I thought this reasonable considering how uncommon it is and how much they have to learn but, when I prompted them further, I was a little 'thrown' when, in answer to the question "What does the word Ectodermal suggest to you?" I drew a blank. They did know what Dysplasia meant.

Unlike the type of medical education I received (and I am nearing retirement) today's students spend little or no time on embryology - the study of the development of the human embryo in the womb. Three basic tissues develop into all body structures - ectoderm, mesoderm, and endoderm. Put simply, the endoderm gives rise to the gut and associated organs, along with the lower respiratory tract (lung tissues); the mesoderm to muscle and skeleton; and ectoderm to the skin, with its associated structures, and nervous tissue, including the brain.

Though generally true, this rule is not hard and fast and you may find, for example, more of an ectodermal contribution to the lungs in one individual than in another. Also, in the formation of the final structure, it is the interplay between these developing components that is the determining factor. For example in certain types of ED there are abnormalities of the hard and soft palate. You wouldn't expect this, these being bone and muscle respectively (and thus of mesodermal origin), but they are covered with tissue of ectodermal origin and it is the mesoderm/ectoderm interplay which leads to the fault. Had they known this, they could have made a good shot as to those tissues, structures and organs in which problems might arise for the individual with Ectodermal Dysplasia. They could have gone further by realising to what structures ectoderm contributes in the ear, nose, and throat, and thus guess what symptoms and problems might come my way as a practising ENT surgeon.

In the ears, nose, and throat ectoderm contributes to the final form and function of these structures:

1. The Ear

- a) The pinna, or aurical, and external ear canal
- b) The nervous tissue of the inner ear - those nerves which pick up and carry sound to the brain

2. The Nose

The mucous membranes (lining) of the nasal cavities and sinuses

3. The Throat

As I mentioned above, ectoderm lines the upper part of the respiratory tree (of which the nose is part), the voice box, and then, variably, the main windpipe and occasionally beyond down toward the lung tissues themselves.

4. The Palate

The palate (roof of the mouth) is part of both the upper respiratory tract, allowing breathing through the nose and speech, and upper gut, where it is an important part of the swallowing mechanism preventing the food and drink you take in ending up in your nose. Not only does a problem here cause speech and swallowing problems, the palate is crucial to the normal function of the ear allowing normal hearing. I will discuss the palate and its role here when considering the ear, as the speech and swallowing problems are more usually the province of the plastic or facio-maxillary surgeon, rather than an ENT surgeon.

Taking these regions in turn:

The Ear

ENT surgeons consider diseases of the ear as falling into three areas. The External Ear (auricle and external ear canal), the Middle Ear (ear drum, ossicles - the small bones in the ear, and the Eustachian tube), and the Inner Ear (the organ of hearing and balance, with the nerves linking them to and into the brain). Let us take these three regions one by one.

The most frequent problems patients with ED encounter centre on the external ear, specifically in the ear canal. The ear canal is lined with skin, which is of course of ectodermal origin, but the skin of the ear canal is unique and differs from the skin covering the whole of the rest of the body. Skin is constantly replacing itself and does so by new cells appearing in the deeper layers. They grow and mature and, as they do so, they move up towards the surface to replace their predecessors who are by then, when on the very surface, dead. The cycle repeats itself endlessly. The dead cells are 'rubbed off' by clothing and physical contact and are lost into the environment. This setup would not work in the ear canal. The dead cells would simply build up as there is nothing there to rub them off and, within a short while after birth, the ear canal would be chock-a-block with dry debris. In the ear canal there is a special mechanism to deal with the normal dead skin. Here the new cells arise on the ear drum and as they mature they move, not from the deep to the surface layer, but out along the ear canal, maturing only when they reach the outer part of the canal where they finally die and can be shed to the outside world.

Early experimenters discovered this by putting Indian ink dots on the ear drums and then watching over subsequent days and weeks. The spot moved relentlessly to the outer ear canal and disappeared as the stained cells were shed into the environment.

Some patients with ED lack this migratory mechanism and their ear canals do indeed become chock-a-block with dead skin to a degree where their hearing is affected - they can't hear through it and, occasionally, the dead skin becomes infected causing inflammation of the ear canal. This is Otitis Externa.

The traditional way to remove wax (and actually wax is largely composed of dead skin sheddings anyway) is by syringing. The problem in ED is usually resistant to this measure (though it may be worth a try). The only answer, and the way I manage those patients with ED and this problem under my care, is to remove the debris from the ear canal under direct vision using an operating microscope and picking, or sucking, it out piecemeal. Before doing it, I ask the patients to put olive oil in the ear, a few drops twice or three times a day for the day before cleaning (aural toilet), as this softens things up and also lubricates the canal walls allowing me to extract even quite hard lumps painlessly. It goes without saying, no patient, and not only those with ED, should attempt to winkle wax out of their own ears with any sort of gadget. It is easy to damage the ear canal wall and to start infection. (The old adage 'Put nothing in your ear smaller than your elbow' is a good one). I do this perhaps every six months or so for my patients. It varies.

Middle ear problems, when they do occur in ED, are caused only when there is a cleft palate. A crucial part of the middle ear mechanism carrying sound to the inner ear is the tube that connects the middle ear to the upper respiratory tract - the Eustachian tube, which runs from the middle ear to the back of the nose. It keeps the air pressure in the middle ear at the correct level (actually that of the atmosphere) - it ventilates the middle ear. If middle ear pressure falls or rises, the ear drum can't move properly and hearing deteriorates. This tube is constantly opening and closing through the day to make the necessary fine adjustments. The tubes can only function normally if the palate is intact, i.e. joined in the middle, which is of course not the case with a midline cleft. Potentially then those children born with a cleft could run into hearing difficulties if nothing were done (and in the past this was sometimes the case) or, if anything was done, it was not for some years after birth and, by then, middle ear problems had set in. Nowadays however cleft palate surgery is so good and done so soon after birth that only rarely does a cleft palate, whether with or without ED, lead to problems and hearing difficulties. If it does, we can do one of two things:

1. Provide an alternative 'ventilation' route. This is done by putting a tiny ventilation tube, a grommet, in the ear drum to allow pressure to equalise across the drum.
2. Provide the individual with a hearing aid which works extremely well with this type of hearing loss.

Inner ear problems do occur (rarely) amongst the various forms of ED. This sort of hearing loss is referred to loosely as 'nerve' deafness or, more correctly, as 'sensori-neural' hearing loss. When a clinician uses that term he or she means that the fault lies in the

inner ear, in the organ of hearing, or in the nerves linking that organ to the centres of understanding and hearing in the brain.

In truth, little is known about this type of hearing loss in ED. It may involve one ear or both, it may be mild or severe, it may be progressive, i.e. worsening with age. It is very difficult then to say to a particular patient, or that patient's parents, what the future holds in hearing terms. It is likely however that, if the child is not actually born with the loss, it will develop or become apparent early in life. The guiding principle for the doctor must be to recognise the possibility that a child considered to have an ED might, and I repeat might, just have a hearing loss, to look for it right from the word 'go' and establish what level of hearing the child has. Continual monitoring of hearing will have to continue through the child's life.

Right from birth we now have methods of testing a neonate's hearing. Doctors recognise certain categories of children who are 'at risk' of having hearing loss which include such things as prematurity, abnormalities of the head and neck and a family history of hearing loss amongst them. Such children will be 'screened' for possible hearing loss. In time all neonates will ideally be screened. At the moment it is not 'universal' in the British Isles. One of the problems in ED is of course that the condition is often not recognisable, nor recognised at birth, so they may 'slip through the net'.

The problem with cells in the central nervous system is that once destroyed they cannot be replaced. What one must do then is make the most of what cells the individual has remaining, i.e. what level of hearing the patient has and there will always be some. This means diagnosing the condition as early as possible and making proper provision for the child's learning and education which must suit the level of hearing loss. In this form of deafness the mainstay of education will be some form of hearing aid. Simply this amplifies the incoming sound to a level where the impaired inner ear can register it. A child introduced early enough to a hearing aid will readily get used to it and adopt it as part of their lifestyle. The hearing aid, however, is only part of the 'package' needed to allow the child with hearing loss to realise their full potential. Nowadays not only Health Care Professionals - Doctors and Health Visitors, District Nurses etc. - but parents and teachers are alert to hearing loss in children under their care. I repeat that hearing loss is not a common feature of ED.

The Nose and Throat

I will take these two together because the problems that may occur in ED in these areas have a common mechanism - a failure in function of the respiratory epithelium. The lining of the nose and respiratory passages down into the lungs is actually a sophisticated barrier preventing infection gaining access to the body. This epithelium (epithelium meaning simply a covering or lining) has two functional elements which work together. First there are in the epithelium many cells which produce, or secrete, a mucus. The products of these cells coalesce to form a mucus blanket covering the surface. Second, the surface cells have little 'hairs', cilia, on their surfaces projecting into the mucus blanket. These cilia move and they do so in a purposive manner acting together to waft or

push the overlying mucus blanket in one intended direction. In the nose they all pull together directing the mucus blanket towards the back of the nose. In the voice box and below they direct the mucus up and out of the voice box. In both instances the mucus ends up in the throat. Anything potentially harmful you breathe in will thus alight on, and become trapped in, the mucus blanket and this continuous transport system will ensure that the 'contaminated' mucus ends up where it can be swallowed and pass harmlessly through the gut. As this epithelium is of ectodermal origin, both functional elements may be impaired to a degree - in most ED patients hardly, if at all, but occasionally the problem may become extremely troublesome. Deficient mucus production and impaired ciliary activity means local inflammation. Sticky secretions build up and become dried out in the inspired air forming adherent crusts in the nasal cavities and the main passages to the lung tissues. This is certainly unpleasant for the sufferer with a dry and crusty nose and occasionally more worrying when the build-up of crusts in the airways to the lungs threatens to obstruct breathing. What can the ENT surgeon and the patient do to help? We cannot 'replace' the defective lining, but we can help it do its job.

1. We can help what cilia are functioning by making the abnormal sticky mucus, which is hard to move along, runnier or less viscid. Humidification of the inspired air will help to loosen things up and, in the case of the chest, assisted by physiotherapy, shift the thickened mucus and crusts. How often humidification is needed varies greatly between patients throughout the year, depending on atmospheric conditions, and on the working environment, temperature and humidity. Nasal douches of a simple agent such as Sodium Bicarbonate are extremely helpful.
2. Humidification and nasal douches are a problem in children who have, certainly in their own eyes, better things to do and clinicians have to resort to toilet of the nasal cavities and clearance of crusts under a general anaesthetic. Similarly in both children and adults bronchial toilet, again under general anaesthetic, may occasionally be necessary when things get out of hand. With my handful of patients I have not had to do this and I think most ENT surgeons will have similar experience. However, if it is necessary, nasal toilet is no problem for most ENT surgeons and, if an ENT surgeon is not happy with bronchial toilet, then a thoracic surgeon has the necessary skill.

You now, after reading this, will certainly know more than most medical students and indeed most doctors about the ENT problems that may affect patients with ED. Certainly I know two students who do know about it, or rather did, in the clinic. Let's hope they retain something of it.

This article was first published in our newsletter (Volume 2 Issue 1 - November 2001).