

Common Ear, Nose and Throat Problems in Ectodermal Dysplasia

by

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The specialists most often involved in the management of ED are dermatologists (skin specialists) and dentists with a special interest in patients with reduced numbers of teeth. However, some manifestations of ED are likely to lead to a referral to an ear, nose and throat specialist.

Absence or reduced numbers of mucous glands lead to abnormal functions of the linings of the nose, sinuses, Eustachian tubes and throat. The salivary glands may also be small or absent, as may the mucous glands in the entire bronchial tree and gastrointestinal tract. These abnormalities lead to a number of different problems.

People with ED may suffer from a blocked nose and sinusitis. Recurrent nosebleeds may occur because of nasal dryness. The nose frequently contains thick mucous and crusts. Nasal symptoms can be alleviated by the use of steam inhalations and nasal douches. However, there is no treatment which can cure these problems.

For the same reason dry mouth and throat irritation often occur. A number of individuals also have difficulty in swallowing due to dryness of the throat. Avoiding excessively dry foods and adding lubricants such as fruit, water, sauces or custard can facilitate swallowing. Drugs which stimulate the production of saliva and artificial saliva preparations may be of value.

Hoarseness of the voice is usually due to laryngeal dryness and can be alleviated by humidification. In addition, nodules on the vocal cords have been reported. These may improve with speech therapy, but in some cases they need to be removed.

Recurrent upper respiratory infections may occur. These are due to viral infections and are therefore self limiting. Symptomatic treatments, such as paracetamol and steam inhalations, are helpful. Some individuals also experience recurrent ear infections. These are particularly likely in association with a cleft palate. Such episodes can be treated with antibiotics, but they do resolve even if no treatment is given. In cases with cleft palate the presence of fluid in the middle ear is also common and in some cases develop chronic ear infection with perforation of the eardrum. These problems may require surgery.

Some types of ED are associated with inner ear deafness. The narrowness of the ear canal leads to wax impaction that may exacerbate hearing loss due to chronic ear disease or abnormalities of the inner ear. It also makes the fitting of hearing aids more difficult. If the ear canals are very narrow, an operation to make them bigger may be helpful.

In children, the first manifestation of ED may be high fevers, recurrent ear infections, nasal obstruction or difficulty in swallowing. A hoarse cry and severe wax impaction in the ears may also be the first indications of the disorder. ED children become febrile easily in hot environments, because of their inability to sweat.

The recognition of these problems and their association with ED permits early management with a multidisciplinary approach. An individual ear, nose and throat specialist will only see a few patients with ED during his or her career. To inform them about ED and its impact on the ear, nose and throat we have recently published an article in the Journal of Laryngology and Otology which covers all the problems they are likely to see.

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