Treating Ear Problems in Ectodermal Dysplasias



Testing Hearing

If hearing loss is suspected, the affected person should be evaluated. If the affected person is a child, someone experienced in audiometric testing of children should do the evaluation. No child is too young to be tested. In the United States, all newborns are routinely screened for hearing deficits and many schools perform hearing screenings.

Testing Types

If indicated, a variety of age-appropriate testing methods are available. In the young child, the evaluation of hearing should include otoscopy (looking inside the ear), impedance audiometry, and behavioral audiometry. If behavioral testing is not possible or the information is not reliable, brain stem evoked response audiometry ("BAER") should be done. These tests will determine whether or not hearing loss exists and may indicate its cause or guide further testing. If a hearing loss is found, assessment by an ear, nose, and throat (ENT) physician is essential.

Ear Wax Problems

Abnormal wax production or accumulation may be noticed in some people affected by ectodermal dysplasias. This should be expected since wax glands are specialized types of sweat and skin glands. The most frequent problem is excessive accumulation of wax in the ear canal. This may result in severe itching and even hearing loss. If a problem exists, lubricant drops (either mineral oil, glycerin, or a 50/50 mixture of rubbing alcohol and white vinegar) may soften the wax.

If these methods are not effective, the wax can be removed as directed by a physician. Periodic cleaning by a physician may be necessary if the problem is chronic. Cotton swabs (Q-tips) should NOT be used to remove wax from the ear, as they may cause injury or further pack the wax into the ear. Wax can be removed (usually painlessly) through a variety of techniques including flushing the ear canal with water or through the use of a microscope and special instruments.

The content of this document is for informational purposes only. Questions regarding specific patient issues should be directed to the appropriate professionals for resolution.

