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A Study of a Dutch Family with Progressive Heredo-degenerative Perceptive Deafness Comprising 334 Members

A.H.		van Bolhuis
D.W.		Odenthal
E.H.		Huizing

From the Ear, Nose and Throat Department of the University Hospital, Leyden

Authors' address: Dr. A. H. van Bolhuis, Dr. D.W. Odenthal and Dr. E. H. Huizing, E.N.T. Department of the University Hospital, Leyden (Netherlands)

I. Genetic Aspects

A family comprising 334 members in 5 generations with progressive perceptive deafness has been the subject of an extensive study. 67 members were found to be affected by the familiar type of perceptive impairment. The inheritance of the trait was of the simple dominant autosomal type with complete penetrance.

II. Audiological Aspects

By D. W. ODENTHAL, E. H. HUIZING and A. H. VAN BOLHUIS

An analysis of the continuous threshold audiograms of the affected persons showed a systematical way of progression of the impairment. 4 typical stages of degeneration were distinguished on account of the audiometric curves.

The question is raised whether this typical degeneration is related with the recent electrophysiological data, i.e. 1. the difference found between the low and high frequencies in the cochlea and 1st neuron and 2. the difference between inner and outer hair cells.

., * Will be published in extenso in this journal.