Approximately 1 in 750 live births presents with severe-to-profound hearing loss. The incidence of hearing impairment is considerably higher for less severe losses. Although the Joint Committee on Infant Hearing has underscored a series of factors that place an infant at risk for hearing loss, less than one-half of all hearing impaired infants are identified through its implementation. The remainder of infants and children identified with hearing loss result from genetic factors, most often the result of single gene mutations, which present as one of either autosomal dominant, autosomal recessive, or sex-linked traits. Further, there has been an increasing number of infants identified with hearing loss that result from the interaction of environmental factors and gene mutation.

Whereas the majority of the 200 types of hereditary hearing loss occur without clinical indicators, about one-third of all genetic hearing loss accompanies syndromes with recognizable physical characteristics. Generally, it is this latter group that has received the greatest attention and the most study. Because of the nature of their physical signs and symptoms, infants with recognizable syndromes are simply more readily identified and at an earlier age.

Much of the current early identification of genetic disorders has been attributed to recent technological advances in human genetics. As a result, a number of heretofore unknown forms of inherited syndromic disorders have been mapped by genetic-linkage analysis. Newer genetic techniques have been used successfully to locate and characterize genes for several types of hereditary hearing loss. Unfortunately, much remains unknown about the genetic abnormalities of nonsyndromic hearing loss. The effect of this newly acquired knowledge on clinical practice provides a greater opportunity for earlier detection, prevention, and potential treatment.

In the assessment and diagnosis of inherited hearing loss, the majority of medical and other ancillary services tend to be confined within the boundaries of a medical environment. For audiologists who work in nonmedical facilities, it is rare to become directly involved in the diagnosis and management of these patients. This special issue of JAAA, entitled “Hereditary and Other Childhood Auditory Deficits,” is devoted to common and unique inherited and multifactoral auditory disorders.

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