



Article

Congenital Nonprofound Bilateral Sensorineural Hearing Loss in Children: Comprehensive Characterization of Auditory Function and Hearing Aid Benefit

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Abstract: A prospective cross-sectional design was used to characterize congenital bilateral sensorineural hearing loss (SNHL). The underlying material of >30,000 consecutively screened newborns comprised 11 subjects with nonprofound, alleged nonsyndromic, SNHL. Comprehensive audiological testing was performed at ~11 years of age. Results showed symmetrical sigmoid-like median pure-tone thresholds (PTTs) reaching 50–60 dB HL. The congenital SNHL revealed recruitment, increased upward spread of masking, distortion product otoacoustic emission (DPOAE) dependent on PTT (<60 dB HL), reduced auditory brainstem response (ABR) amplitude, and normal magnetic resonance imaging. Unaided recognition of speech in spatially separate competing speech (SCS) deteriorated with increasing uncomfortable loudness level (UCL), plausibly linked to reduced afferent signals. Most subjects demonstrated hearing aid (HA) benefit in a demanding laboratory listening situation. Questionnaires revealed HA benefit in real-world listening situations. This functional characterization should be important for the outline of clinical guidelines. The distinct relationship between DPOAE and PTT, up to the theoretical limit of cochlear amplification, and the low ABR amplitude remain to be elucidated. The significant relation between UCL and SCS has implications for HA-fitting. The fitting of HAs based on causes, mechanisms, and functional characterization of the SNHL may be an individualized intervention approach and deserves future research.

Keywords: children; congenital; electrophysiology; hearing; hearing aids; imaging; otoacoustic emissions; sensorineural hearing loss; speech recognition

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1. Introduction

The introduction of universal newborn hearing-screening (UNHS) has changed pediatric audiology substantially—like a “silent revolution” [1]. While the UNHS [2–4] and early intervention with hearing aids (HAs) seem to be beneficial (e.g., [5–7]), and cost-effective [3,8], early diagnostics and intervention are indeed challenging. The current lack of comprehensive auditory characterization of typical congenital sensorineural hearing loss (SNHL) hampers further advances in early intervention. More knowledge is needed on the causes (see recent reviews on etiological evaluation [9,10]), mechanisms, and the resulting dysfunction, to enhance subsequent communicative capacity.

The long-term goal is to find the causes and mechanisms behind various nonsyndromic congenital SNHLs and to identify or develop future treatment options. Here, we try to characterize the summary auditory profile and potential mechanisms of alleged nonsyndromic, nonprofound, bilateral congenital SNHL. A comprehensive audiological

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