



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

Hearing Lob In Children A Comprehensive Text

Daniel Tranel, Paul J. Eslinger



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Hearing Loss in Children Burton F. Jaffe,1977 *Hearings on Child Care* United States. Congress. House. Committee on Education and Labor,1989 *The Oxford Handbook of Deaf Studies in Literacy* Susan R. Easterbrooks,Hannah M. Dostal,2021 The Oxford Handbook of Deaf Studies in Literacy brings together state of the art research on literacy learning among deaf and hard of hearing learners DHH With contributions from experts in the field this volume covers topics such as the importance of language and cognition phonological or orthographic awareness morphosyntactic and vocabulary understanding reading comprehension and classroom engagement written language and learning among challenged populations Avoiding sweeping generalizations about DHH readers that overlook varied experiences this volume takes a nuanced approach providing readers with the research to help DHH students gain competence in reading comprehension

The Complete Handbook of Children's Reading Disorders Hilde L. Mosse,1987 M D Hilde L Mosse head psychiatrist for the N Y City Board of Education writes from her own highly successful experience in teaching 1000 psychologically disturbed 503 646 9459 *Politzer's Text-book of the Diseases of the Ear for Students and Practitioners* Adam Politzer,1926

A Text-book of pathology systematic & practical v.2 pt.2, 1894 David James Hamilton,1894 Text Book Of General Physiology P.V. Jabde,2005 Physiology as such a vast subject with unlimited scope and which can be studied in different ways There was a long left need among students and teachers for a text book with recent information and written according to national syllabus Efforts have been made to collect recent informations from several standard works and journals Contents Nutrition Circulation Respiration Physiology of Aviation High Altitude Space and Deep sea Diving Thermo Regulation Osmoregulation Excretion Muscles Nervous System Reproduction Sense Organs Chemical Messengers Physiology of Endocrine Glands Chronobiology **Illustrated Text Book of Neuroanatomy** GP Pal,2013-01-01 Illustrated Textbook of Neuroanatomy Presents a comprehensive yet lucid and friendly coverage of neuroanatomy explains the concepts in a simple and easy to understand language **Early Frontal Lobe Damage and Development** Daniel Tranel,Paul J.

Eslinger,2001-05 The consequences of early onset brain damage for the development of cognition and behavior have recently been identified as top research priorities by the NINDS This special issue presents a series of new empirical studies that address this issue in depth from several different perspectives and in both human and animal participants The focus is on the development of personality social behavior and related executive functions in subjects who suffered early damage to prefrontal brain regions A consistent theme throughout is that early onset prefrontal lesions can frequently lead to severe intractable deficits in social behavior and moral reasoning and to impairments of executive functions such as planning judgment and decision making *A Text-book of Pathology* David James Hamilton,1894 **A Text-book of Medical Practice for Practitioners and Students** William Bain,1904 *A Treatise on the Nervous Diseases of Children* Bernard Sachs,1895 *A Text-book of the Diseases of the Ear* Josef Gruber,1893 *The Works of William Shakspeare, Complete.*

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