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Epileptiform electroencephalogram abnormality in children with congenital sensorineural hearing loss

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ABSTRACT

Objectives: This work was designed to study electroencephalogram findings in children with congenital sensorineural hearing loss and correlate these findings with the SNHL parameters as duration, etiology, severity, and type.
Methods: Ninety children with bilateral congenital sensorineural hearing loss served as the study group. They were free from any neurological disorders or symptoms that are commonly associated with abnormal electroencephalogram as convulsions or loss of consciousness. Twenty children having normal hearing with no history of otological or neurological disorders served as the control group. All children participating in the study were subjected to full medical and audiological history, otological examination, neurological examination, audiological evaluation and electroencephalogram recording.
Results: Mean age of the children in the control group was 3.56 ± 2.1 years and mean age of the children in the study group was 3.8 ± 2.2 years. While none of the control children had abnormal electroencephalogram, 38 (42.2%) of children with congenital SNHL had epileptiform electroencephalogram abnormality. The epileptiform abnormality was generalized in 14 children (36.8%), focal temporal in 17 children (44.7%) and focal other than temporal in 7 children (18.4%). According to the hemispheric side affected, the abnormality was right in 14 children (36.8%), left in 10 children (26.3%) and bilateral in 14 children (36.8%). No statistically significant predominance of specific site or side of the epileptiform abnormality was found. Similarly, no statistical significant prevalent of the epileptiform abnormality was found in relation to the age or sex of children, duration of hearing loss or etiology of hearing loss (i.e., genetic vs. neonatal insults). On the other hand, the epileptiform abnormality was statistically prevalent in children with moderate degree of hearing loss, and in children with auditory neuropathy spectrum disorder.
Conclusions: The epileptiform electroencephalogram abnormality is a common finding in children with congenital sensorineural hearing loss especially those with auditory neuropathy spectrum disorder, suggesting the affection of the central nervous system despite the absence of neurological symptoms or signs. These findings raise the question of the requirement of medical treatment for those children and the effect of such treatment in their rehabilitation.

1. Introduction

Congenital sensorineural hearing loss (SNHL) is one of the most common congenital sensory deficits [1]. Universal neonatal hearing screening programs in developed countries revealed an incidence for the congenital SNHL of 2-4 neonates per 1000 neonates [2,3]. The incidence of congenital SNHL in the developing countries is likely to be much higher [4,5]. If left untreated, the implications of congenital SNHL are drastic for the children and their families. These implications include delayed language development, scholastic under-achievement, behavioral, psychological, and social problems [6]. Fortunately, these implications can be prevented or severely minimized with early identification and early intervention with the appropriate and adequate rehabilitation program, including fitting with suitable amplification device either hearing aids or cochlear implant [7,8].

In most cases, the pathology of congenital SNHL is essentially in the cochlea (i.e., cochlear hair cells, steriocilia, striavascularis, and/or tectorial membrane). Auditory neuropathy spectrum disorder (ANSD) is the term suggested by the international panel of experts in Como, Italy (2008) [9] to describe the hearing disorder that was first labeled by Starr et al. [10] as auditory neuropathy. ANSD is a frequent presentation in pediatric population with SNHL, accounting between 10% and 14% of children with SNHL [11,12]. The prevalence is much higher in hearing impaired infants graduate from the neonatal intensive care unit or have risk factors, whereas

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40–77% of those infants were reported to have ANSD [13,14]. While the pathological site of congenital SNHL can be classified as cochlear in the majority of cases, the pathological site in children with ANSD can be classified as neural whereas the cochlear outer hair cells (OHCs) are functioning well (as shown from the intact otoacoustic emissions (OAEs) and cochlear microphonics (CM)) while the neural function are severely disrupted (as shown from absent or severely abnormal compound action potential (CAP), and auditory brainstem response (ABR) starting from wave I) [10]. Rather than single pathological site, several pathological sites underlying ANSD were suggested. Theses sites include the synapse between the inner hair cells (IHCs) and auditory nerve afferents, distal and proximal parts of the auditory nerve, and higher auditory nervous system [15].

In the vast majority of children, the onset of ANSD occurs before language development leading marked language impairment [16,17]. As regards language acquisition, hearing aid (HA) was reported to be successful only in about 15% of children with ANSD, while cochlear implantation (CI) was successful in more than 80% [17]. Proper diagnosis of the site of lesion is important in the prognosis of children with ANSD and, in the choice of amplification device (HA vs. CI) and rehabilitative methods.

Electroencephalogram (EEG) provides measures of cortical function with excellent time resolution. It is especially valuable in investigation of patients with known or suspected epileptic seizures. The mainstay of diagnosis of epilepsy in children is detecting interictal epileptiform EEG pattern. Such epileptiform EEG pattern is rarely seen in the EEGs of non-epileptic children [18]. The prevalence of epileptiform EEG pattern among normal child population was reported to be between 0.4 and 1.25% [19]. However, it has been our clinical observation that children with congenital SNHL frequently demonstrate an epileptiform EEG pattern in the absence of history or current epileptic fits. Upon reviewing the literature, only scarce and old studies on EEG findings in congenital SNHL were found. Zwirecki et al. [20] found that 16 out of 25 (64%) deaf children demonstrated epileptogenic focus in EEG recording. Fishman et al. [21] found that 17 out 31 (54.8%) children in a deaf school had abnormal EEG patterns; 6 of these children had epileptogenic record.

In human subjects, the epileptiform EEG pattern is the electroencephalographic hallmark of interictal recording in epileptic patients [22]. It reflects hypersensitivity at the level of the cerebral cortex. Thus the excitatory influences outweigh the inhibitory influences [23]. Considering the reported high prevalence of EEG abnormalities in congenital SNHL [20,21] and our clinical observation that children with congenital SNHL frequently demonstrate an epileptiform EEG abnormality in the absence of history or current epileptic fits, we hypothesize that congenital SNHL raises the excitability within the cerebral cortex leading to the epileptiform abnormality in the EEG recording.

The current work was designed to study EEG findings in children with congenital SNHL in order to determine the prevalence of EEG epileptiform abnormality in children with congenital SNHL and correlate EEG findings with SNHL parameters as duration, severity, etiology, and more specifically the type (i.e., cochlear vs. ANSD). Results of the current study may add new information about the pathogenesis of SNHL specially the involvement of the CNS. Moreover and clinically relevant, proper management of the abnormal EEG in children with congenital SNHL may have positive effects in the rehabilitation process of those children.

2. Subjects and methods

The current study consisted of 110 children classified into study group and control group. The study group consisted of 90 children with bilateral congenital SNHL. Children of the study group were recruited from children attending the Audiology clinic in Minia university hospital, Minia, Egypt. They were just diagnosed with bilateral SNHL and not fitted with hearing aids yet. Inclusion criteria were bilateral congenital SNHL hearing loss and absence of any neurological disorder or symptoms that are commonly associated with abnormal EEG as convulsions, loss of consciousness or behavioral complains. The control group consisted of 20 children selected to be age and sex matched with the study group. They were selected from children of medical staff members, and relatives accompanying patients in the Audiology unit, Minia university hospitals. All children in this group had no history of otological or neurological disorders. They had bilateral normal hearing, and normal speech and language development. For both study and control groups, written consents were taken from the parents for their children participation after detailed explanation of the study procedures. All procedures were approved by the ethical research committee in Minia University, Egypt.

Age of children in the study group (90 children) ranged from 1 year to 13 years with mean age of 3.8 years and SD of 2.2 years. They were 50 boys (55.6%) and 40 girls (44.4%). Age of the children in control group (20 children) ranged from1 year to 11 years with mean age of 3.56 years and SD of 2.1 years. They were 11 boys (55%) and 9 girls (45%). No statistical significant difference was found between the study group and the control group as regards the age (t value = 0.2; p value = 0.83) or gender distribution. (Chi square value = 0.2; p value = 0.66).

All children in the study group had bilateral congenital SNHL; 16 (17.8%) of them had ANSD. Reliable audiometric threshold could not be obtained for 6 children with ANSD. In the rest of children (number = 84) reliable audiometric threshold was obtained through either behavioral audiometry alone or combining results of behavioral audiometry with ABR results. Six children (7.1%) had mild hearing loss, 26 children (31%) had moderate hearing loss, and the majority (52 children; 61.9%) had severe or profound hearing loss.

All children in the study group had congenital SNHL in the sense that had the hearing loss before speech and language development. Based on the history, genetic causes were suspected in 33 children (36.7%), and neonatal insults were the causes in 35 children (38.9%). In the remaining children (22 children; 24.4%), the causes were unknown. Neonatal insults included hyperbilirubinemia in 11 children (31.4%), low birth weight (LBW) in 7 children (20%), hypoxia in 6 children (17.1%), premature labor in 6 children (17.1%), and fever in 5 children (14.3%).

All children participating in the study were subjected to history taking, otological examination, neurological examination, audiological evaluation, and EEG recording. History included full medical and audiological history. It included the full description of prenatal, perinatal, neonatal, postnatal, developmental and family history, in addition to the neurological history specially history for epileptic fits or loss of consciousness. Basic audiological evaluation included immittancemetry (tympanometry and acoustic reflex threshold), and audiometry to assess hearing sensitivity. According to the child age, either behavioral observation audiometry (BOA), visual reinforcement audiometry (VRA), or pure tone audiometry (conditioned or conventional audiomtery) was performed. Immittancemetry was done using the middle ear analyzer Zodiac 901. Pure tone audiometry was done using Audiometer Amplaid 309. For infants, children younger than 3 years, and children in whom reliable behavioral thresholds could no be obtained, ABR was done to estimate hearing sensitivity. In cases showing absent or severely abnormal ABR, the CM was recorded to diagnose ANSD. Criteria for diagnosis of ANSD were absent or severely abnormal ABR with intact CM and absent acoustic reflex [17].