

Otoacoustic emissions: a new method for newborn hearing screening

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Abstract. – Pediatric deafness is a handicap affecting approximately 2/1000 newborns. Currently, its diagnosis is markedly delayed, since it occurs approximately at 24 to 36 months of age; at this age rehabilitation procedures (i.e., acoustic prosthesis, speech therapy, psychological interventions on the family, or cochlear implants in the most serious situations) are unable to ensure a complete development of both the voice and the speech, thus preventing the full participation of the deaf child in social living. The turning point has taken place when methods and techniques were developed; they are aimed at the very early diagnosis of infantile deafness and are based on the recordings of otoacoustic emissions, that is, acoustic signals of extremely weak intensity originating in the inner ear, which not only is a passive transducer, but is able to generate sounds also. Any lack of or any change in otoacoustic emissions is an accurate index of disabling deafness. The test under study allows to perform selectively a mass screening on newborns (it is carried out 2 or 3 days after birth) since it is definitely non-invasive, it is done very rapidly (a few seconds only), it is cost-effective and highly reliable. The newborn hearing screening is being accepted, at a faster growing pace, by an increasing number of health systems in the whole world.

Key Words:

Otoacoustic emission, Distortion Product OtoAcoustic Emissions (DPOAEs).

Introduction

The pathogenesis of pediatric deafness is quite variable and often unknown. However, the prevalence of this auditory handicap in the infantile population is approximately 2/1000 infants¹. Severe and/or marked sen-

sorineural deafness due to genetic or congenital cause affects 1-2/1000 healthy newborns and involves from 4% to 5%^{3,4} of newborns who show at birth one or more audiologic risk factors^{2,5}. At any rate, the prevalence of infant deafness is higher than that of other congenital diseases, such as phenylketonuria and hypothyroidism, for which newborns are routinely submitted to screening procedures. The period of "cerebral plasticity" is established in the first 3 years of life, during which complex and organized interneuronal circuits are developed. Peripheral acoustic input is essential for the proper maturity of central auditory pathways and allows to acquire both auditory memory and speech^{6,7}. The deaf child has no chance to develop adequately a normal ability to concentrate and pay attention. As a consequence, speech reception and expression are limited, leading, in turn, to changes in emotional maturity, relational difficulties, and sociocultural integration^{8,9}. In view of such considerations, the top priority goal is the early discovery of genetic and/or congenital deafness. Currently, the mean age of identification of pediatric deafness is still rather high, being established at around 24-30 months¹⁰. The priority of an early diagnosis, aimed at a prompt intervention strategy capable of operating during the period of cerebral plasticity, led the scientific community to multiply the efforts towards the definition of an effective program of newborn hearing screening. It is common knowledge that approximately 50% of identified deafness in infants shows none of the 10 audiologic risk factors indicated by the Joint Committee on Infant Hearing Screening Assessment^{11,12}. Therefore, a hearing screening performed on the so called "audiologic risk" newborns only, implies the missed identification of about

one-half of neonatal deafness. During the last few years, a number of studies¹³⁻²⁰ have demonstrated the possibility of developing a neonatal audiological screening based on the recordings of evoked otoacoustic emissions (EOAEs). Evoked otoacoustic emissions are acoustic signals of non-linear type delivered by the external ciliated cells (ECCs) of the cochlea. Such cells are present in 100% of normally hearing individuals and are the expression of a normal cochlear function; they are stable, reproducible, influenced by all the cochlear nociceptive factors, and absent in hypoacusia higher than 40 decibels.

Depending on the presence or absence of an external stimulation, otoacoustic emissions (OAEs) may be classified in two main categories: spontaneous OAEs and evoked OAEs.

A) *Spontaneous OAEs* (SOAEs) are obtained without any sound stimulation. They are narrow band signals, single or more often multiple, unilateral or bilateral. They can be measured in 40% to 70% of normal ears^{21,22}, and show a sinusoidal shape similar to the typical shape of pure tones. In the majority of the instances they are found in a frequency range from 1 to 2 KHz; it has been supposed, therefore, that they originate from the intermediate-apical portion of the cochlea. Their origin has been ascribed to a double mechanism: a physiologic mechanism^{23,24} linked both to the spontaneous activity of the ECCs and to the activity chemically and/or electrically induced under steady control of the olivocochlear bundle. The second mechanism, indicated as pathologic²⁵, would be related to a sharply delimited "focal" change or damage of the ECCs, whereas in this situation the producers of SOAEs would be the intact cells adjoining the damaged areas. Both the conditions of the test and the characteristics of the measuring instrumentation used influence, to a significant extent, the stability of the intensity level.

B) *EOAEs* are subdivided into the following categories, depending on the characteristics of the administered stimulus:

- *Simultaneous EOAEs* (*SEOAEs*) obtained by means of continuous sonorous stimuli;
- *Transient EOAEs* (*TEOAEs*) obtained by means of clicks or tone-bursts, and better known as cochlear echoes, or Kemp echoes, or transient echoes;

- *Distortion products OAEs* (*DPOAEs*) obtained by the contemporaneous presentation of 2 sound stimuli (f1 and f2), or primary tones, bound together by a frequency relationship; these emissions consist of different frequencies with respect to applied primary stimuli resulting from the combination of f1 and f2 as a result of either their difference or their summation.

Out of these 3 categories of otoacoustic emissions, the SEOAEs, even though are present in around 94% of normal hearing subjects, are the least used in clinical practice in view of the complexity of their recording.

TEOAEs are the best known otoacoustic emissions (Figure 1); in their behalf a wide field of clinical application was quickly gained as early as the 1980s.

Morphologic stability and reproducibility are the main features of these responses^{26,27}, even though great differences may be found among individuals as well as between the ears of the same individual; often the contralateral ear shows a rather similar response pattern. These signals, in normal hearing ears, are present in 98% to 100% of the cases, independently of sex and age^{29,30}.

DPOAEs (Figure 2) are the result of an active process of intermodulation at cochlear level mediated by the ECCs and are obtained by simultaneously signaling two stimulating pure external tones, called primary, namely f1 and f2; the former is deemed to be an external tone whose frequencies are lower than those of the latter.

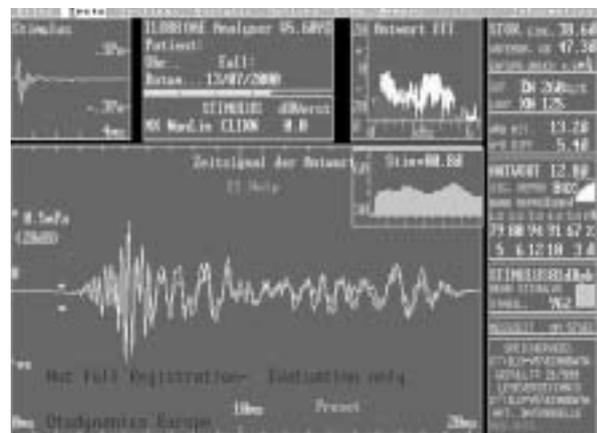


Figure 1. Transient evoked otoacoustic emissions (TEOAEs).

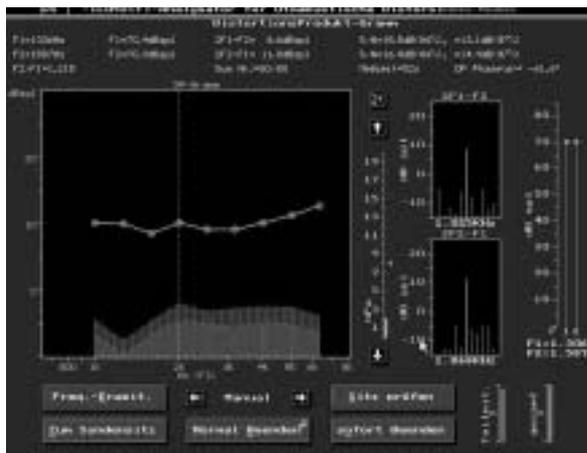


Figure 2. Distortion Products Otoacoustic Emissions (DPOAEs).

Commonly studied DPOAEs correspond to the so called “tones of cubic difference” resulting from the combination $2f_1 - f_2$ ^{31,32}. In fact, they represent the distortion product of the highest intensity in both humans and animals, one of the most stable and certainly that which is linked to the active non linear micromechanisms of the cochlea. Such a feature makes such a distortion product particularly sensitive and quite early vulnerable with respect to the harmful stimuli of toxic, traumatic and degenerative type involving the cochlea. By properly varying the frequency values of either primary tone it is possible to obtain combinations of different frequency; as a consequence, different cochlear areas, both towards the apex and towards the base, may be explored objectively.

DPOAEs can be evoked in over 98% of normal ears. As opposed to TEOAEs, their typical feature is a wide dynamic field and may be recorded even in instances of moderately severe (about 40 to 50 dbHL) sensorineural losses. In contrast with TEOAEs, DPOAEs possess a considerably higher frequency specificity. Therefore, a significant correlation exists between audiometric tonal threshold and DPOAEs' magnitude. Their detection threshold is in inverse proportion to the background noise and is dependent upon the sensitivity of the instrumentation used; under favourable test conditions it is similar to psychoacoustic sensitivity. As far as uses and applications in clinical practice of

DPOAE responses and of TEOAEs are concerned, it is appropriate to specify and emphasize the importance of the middle ear; in fact, the cochlea produces an acoustic emission of very low intensity and therefore a significant otoacoustic emission measured in the ear canal is obtained as a result of a retrograde vibration of the ear drum through the auditory ossicles chain. As a consequence, from what has been said above it can be deduced that only a healthy middle ear may be able to acoustically coupling, in an effective way, the cochlea with the air of the ear canal; thus a normal function of the middle ear is essential for the recording of normal otoacoustic emissions.

Evoked otoacoustic emissions, with particular reference to TEOAEs and DPOAEs, must be considered by now as a powerful means for study and a valuable clinical test for audiological pathological conditions. Otoacoustic emissions are the only specific method of investigation, for the direct and objective study of cochlear mechanisms and dynamics. In general, otoacoustic emissions are simple to perform, absolutely non-invasive and non-traumatic and thus can be carried out in newborns during sleep; they are well accepted by grown up children. The limited cost of the instrumentation used as well as the short time required to perform an examination are additional items on behalf of this diagnostic method.

Among the international experiences, that are points of reference for the use of TEOAEs as a method for newborn hearing screening, we would like to mention the following conferences:

- NIH Consensus Conference in Bethesda (1993), where the Rhode Island Hearing Assessment Project was presented. This is a pilot project started for verifying the validity and the feasibility as well as the costs of an universal screening program to be carried out by using the TEOAEs method.
- European Consensus Development Conference on Neonatal Hearing Screening (1998), where the European Advanced Hearing Assessment Methods and Device Project (AHEAD) were presented; they report the guiding experiences in Europe³³.

TEOAEs have been clinically applied for verifying the presence of a normal hearing function in the newborn and for evidencing simulation phenomena.

TEOAE and DPOAE recordings as screening tests for congenital or acquired auditory defects in infancy have become a reality: a plain and objective diagnostic test for establishing the diagnosis of normal neonatal hearing. Nowadays it is thus possible to examine all the children both at birth and at any subsequent properly selected time period; by doing so, a high qualitative level of audiologic prevention may be achieved. The main problems encountered in the practical application of TEOAEs in the clinical neonatal field are represented by the environmental noise, the endurance of the probe, the presence of a pathologic tubotympanic condition, if any; the latter ailment is a hardly encountered disorder in newborns.

The test under study has shown a high sensitivity, that is, the ability to bring into evidence cochlear deafness in 100% of the cases studied, and a similarly adequate specificity, that is, the ability to sort out normal hearing in 75% to 85% of the cases tested.

The only drawback reported in TEOAE test screening is its low specificity when the cochlear sensorineural pathologic conditions only are the test target. Actually, otoacoustic emissions are found both in normal hearing ears and in ears with retrocochlear or central deafness. In this regard a lively interest has been aroused by recent studies on "auditory neuropathy", whose diagnosis is really based on a normal OAE response against an abnormal Auditory Brain Response (ABR) response. In sensorineural deafness of the adult, TEOAE contribution appears to be limited to the differential diagnosis between cochlear and retrocochlear pathologic conditions, when the lesion does not interfere, at any rate, with cochlear function. TEOAEs may provide some useful hint in subclinical cochlear lesions, in which subjective impairments exist, such as, for instance, acoustic hallucinations and fullness, without any significant change in tonal threshold.

Owing to their features, DPOAEs, in addition to an advantageous use in basic research on physiologic cochlear mechanisms, provide a real possibility of being employed in the study of clinical and preclinical situations of

auditory deficit. From the standpoint of audiometric testing, they appear to be even more interesting, inasmuch as they add, to their features of objective test and non invasive recording, the possibility to select the frequency definition up to the level of fine structure (1 octave) or even of microstructure (0.1 octave), thus allowing an extremely selective mapping of cochlear activity. Moreover, DPOAE study by means of presently available software allows for the analysis of the important functions of increase in cochlear response (input output).

This method starts to be introduced into neonatal screening protocols as an alternative to TEOAEs or in association with them and with a double-check function. In infantile audiometry it may represent a valid complementary diagnostic support to conditioned audiometry and to evoked potentials.

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