B-ENT, 2009, **5**, 79-82

Hearing evaluation in newborns with congenital aural malformation

S. G. Korres,***, D. G. Balatsouras**, C. P. Lyra***, G. S. Korres***, I. Kantas**** and C. E. Papadakis******

*ENT Department of Athens National University, Hippokration Hospital, Athens, Greece; **ENT Department of Tzanion General Hospital, Piraeus, Greece; ***Pediatric Audiology Department, Iaso Maternity Hospital, Maroussi, Greece; ****ENT Department of "G.Genimmatas" General Hospital, Thessaloniki, Greece; *****ENT Department of Chania General Hospital, Chania, Crete, Greece

Key-words. Auditory brainstem response; aural atresia; congenital disease; transiently evoked otoacoustic emissions; universal newborn hearing screening

Abstract. *Hearing evaluation in newborns with congenital aural malformation. Objectives*: Over 5 years, 68,472 newborns were screened through a newly implemented universal newborn hearing screening program. In 15 cases, atresia of the external ear canal was found. The aim of this study was to estimate the hearing status of these newborns using transiently evoked otoacoustic emissions (TEOAEs) and auditory brainstem responses (ABRs).
Methods: TEOAEs were performed during the first days after birth in the normal ears of all newborns. Diagnostic ABR audiometry was performed in 10 newborns.
Results: Unilateral involvement occurred in all newborns studied. TEOAEs were present in all the contralateral ears. Normal ABRs were recorded from the healthy ear, whereas a conductive hearing loss, of approximately 50-60 dB, was found in the involved ear. In long-term follow-up, a satisfactory level of hearing, language, and speech development was found in 9 of the newborns; it was too early to come to definite conclusions in 4 newborns and 2 newborns were lost in follow-up.
Conclusions: In this study, all newborns with aural atresia had normal function in the contralateral ear. Yet, a comprehensive assessment of hearing is essential as early intervention is necessary in the rare case of bilateral hearing impairment.

Introduction

Congenital aural atresia is a birth defect, which is almost always accompanied by abnormalities of both the middle and external ear in varying degrees. It is combined with an inner ear abnormality in approximately 10 per cent of cases.[1] A prevalence of 1 in 3,300 to 10,000 births has been reported.[2] For unknown reasons, it occurs predominantly in males and in the right ear.[3]

The aim of this study was to estimate the hearing status of newborns who presented with aural atresia using transiently evoked otoacoustic emissions (TEOAEs) and auditory brainstem responses (ABRs). This screening was completed through a universal neonatal hearing screening program.

Materials and methods

The study was performed in 'Iaso' maternity hospital, during the implementation of a newborn hearing screening program; the program was initially implemented on a pilot basis and then universally. All newborns with aural atresia born in the past 5 years were included in the study. A detailed anamnestic history was taken from parents, which included questions about a positive family history for congenital malformations involving the ear or other organ sites, as well as use of drugs and alcohol during pregnancy. TEOAEs were performed during the first days after birth on the normal ear of all newborns. Screening was performed on the second day of life and those who failed underwent repeat testing before hospital discharge. Diagnostic ABR audiometry was performed in 10 newborns during the first month of life.

Testing was performed by two ENT specialists experienced in neonatal screening techniques. For TEOAE screening, an ILO88 Otodynamics analyzer (Otodynamics, London, software version 3.94 H) connected to a portable personal computer was used. Details of this procedure are described elsewhere.[4] The ABRs were recorded by a Cadwell Sierra or a Biologic Traveler's system. The stimuli were 100-μs square wave condensation clicks presented with earphones at a repetition rate of 20 Hz. A stimulus level of 80 dB nHL was used initially, 2000 responses were averaged and

the average response wave was replicated twice. Then, the level of the stimulus was decreased in 10 dB steps, down to the threshold of wave V. The lowest stimulus level at which a satisfactory waveform of wave V, with a high correlation between the replications was observed, was defined as the ABR threshold. Right and left ears were stimulated separately, using white noise to mask the nontest ear. Bone conduction was evaluated by presenting bone-conducted clicks at an initial level of 60 dB nHL and progressively decreasing intensities in 10 or 20 dB steps, down to the threshold of wave V. The stimuli were 100-μs square wave alternating clicks presented with a bone oscillator placed at the mastoid, at a repetition rate of 20 Hz. Masking with broadband noise was used in bone conduction testing.

Speech and language development was evaluated by speech and language therapists who employed individualized, tailor-made tests due to a lack of standardized speech and language tests in Greek. Duration of follow-up ranged from 6 months to 3 years, evaluations occurred every 6 months.

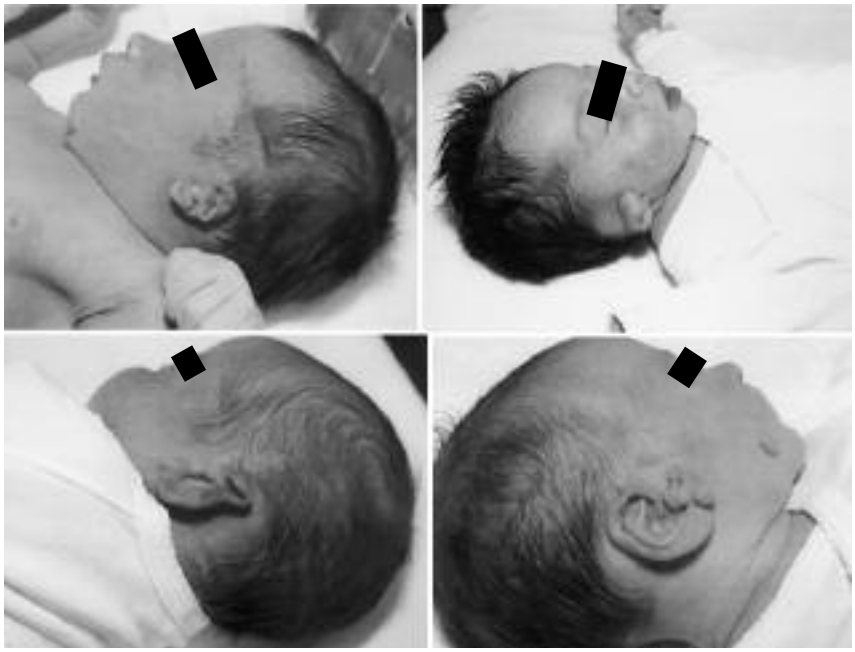

Figure 1
Newborns with aural atresia and malformations of the auricle

Results

Over 5 years of a universal newborn hearing screening program at 'Iaso' maternity hospital, 68,472 newborns were screened and 15 newborns with atresia of the external ear canal were born, with an estimated prevalence of 1 per 4,500 births. The affected newborns included eight males and 7 females. In all newborns, involvement was unilateral with 11 (73%) cases right-sided and 4 (27%) left-sided. Family history was clear in all cases and no risk factors, including drug and alcohol use during pregnancy, were reported. Several newborns with atresia of the external ear and malformation of the auricle are shown in Figure 1.

Congenital disorders or other health problems were absent in 13 newborns. In 1 newborn, renal distention was found that resolved spontaneously after 6 months. A craniofacial malformation was evident in 1 newborn, presenting as a hemifacial microsomia on the side of the involved ear. Computed tomography showed absence of middle and inner ear on the involved side. Two more newborns underwent magnetic resonance imaging, which showed normal middle and inner ear.

TEOAEs were present in all the contralateral ears (Table 1). Normal ABRs were recorded from the healthy ear, whereas a conductive hearing loss, of approximately 50-60 dB, was found in the involved ear. The exception was the newborn with an absence of the inner ear, in whom ABRs were absent on the involved side. In long-term follow-up, a satisfactory level of hearing, language and speech development was found in 9 of the newborns. It was too early to come to definite conclusions in 4 cases and 2 newborns were lost in follow-up.

Discussion

Congenital aural atresia is a relatively common unilateral or bilateral malformation. In our screening, a prevalence of 1 in 4,500 births was found, which is among the higher reported rates of this disorder.[2] We confirmed the predominant involvement of the right versus left ear,[3] but no significant preponderance of males over females was found. Although bilateral involvement has been

Table 1

Mean values (m) of reproducibility scores and response levels of transiently evoked otoacoustic emissions, in the normal ears of newborns with aural atresia. Standard deviation (sd) and range (r) are provided

Reproducibility (%)						
	1 kHz	2 kHz	3 kHz	4 kHz	5 kHz	Total
m	13.4	78.5	86.1	87.7	52.8	67.7
sd	22.9	15.2	16.0	9.4	97.0	14.0
r	0-49	55-92	51-97	73-99	95-99	51-89
Response level (dB)						
m	0.0	6.2	9.7	12.2	13.1	16.1
sd	0.0	4.0	4.9	6.4	6.6	8.4
r	0-0	1-11	0-15	4-23	4-21	2.9-27.2

reported in approximately 9% of all patients,[5] all our cases were unilateral.

Quite frequently, aural atresia is associated with abnormalities of other organs as a result of genetic disorders, chromosomal defects, intrauterine infections, or environmental teratogens.[6] In our patients, only two cases were found to have such abnormalities; one with a mild self-limited kidney disorder and the second with a syndrome of the first two branchial arches characterized by asymmetric face, hemifacial distortion, and hypoplasia.[7] Concurrent involvement of the external, middle, and inner ear is quite common. Several classification systems for developmental anomalies of the ear have been proposed, depending on the degree of malformation and the structures involved. According to the 3-group classification system first proposed by Altmann[8] in 1955, which has been widely accepted with minor modifications,[2] most of the ears of our study belong to the second group of moderate malformations. The ear of the newborn with hemifacial microsomia belongs to the third group of severe malformations, as computed tomography revealed aplasia of the middle and inner ear. However, it should be noted that radiological evaluation was not performed in most newborns and probably one or more ears could be classified into the third group.

In newborns with bilateral involvement, a serious handicap of hearing impairment is present. Fortunately, in most cases unilateral involvement occurs and the other ear has normal hearing. The hearing deficit is of secondary importance and no hearing aid is needed. In any case, a comprehensive assessment of hearing is necessary whether the malformation is unilateral or bilateral. The physician should never assume a newborn with an ear malformation limited to one side has normal hearing on the contralateral ear.

Fortunately, the cochlear status of the normal ear may now be obtained through evoked otoacoustic emissions. This test is becoming one of the routine audiological procedures used to study peripheral auditory dysfunction in a non-invasive and objective manner.[9,10] Evoked otoacoustic emissions can be reliably measured from nearly all newborn ears, if cochlear function is normal and the middle ear transmission system is not defective.[11] In our study, all newborn ears produced normal otoacoustic emissions, indicating newborns with unilateral atresia had normal hearing in the contralateral ear. No immediate treatment was necessary. At the age of 5-6, the option of surgical intervention should be considered. However, atresia repair surgery should be based on stringent audiological and radiological criteria and only be performed by surgeons highly experienced in this type of surgery. Our results are in agreement with a recent study by Suutarla *et al.*,[12] who performed a retrospective review of 190 patients with microtia. In 93% of this group, microtia was associated with atresia or stenosis of the external ear canal, accompanied by conductive hearing loss in most of the involved ears. Sensorineural hearing loss occurred in less than 10% of the group. However, none of the examined patients had hearing loss in the non-affected ear.

Despite normal hearing in the non-involved ear, as detected by the highly sensitive method of otoacoustic emissions, further confirmation by ABR should be pursued.[13] The advantage of ABR is its ability to determine the presence or absence of cochlear function in both ears. Both air and bone conduction thresholds may be obtained and an accurate estimation of the existing air-bone gap of the involved ear may be provided. In all newborns examined in this study, normal ABRs were recorded from the healthy ear, whereas an air-bone gap approximating 50-60 dB was found in the involved ear of most.

Early determination of cochlear function is critical. In cases of bilateral impairment, auditory training should begin during the first year of life to achieve normal speech and language development.

Conclusions

The following may be concluded from this study:

- TEOAEs, in conjunction with ABRs, may help to estimate the cochlear status of the uninvolved ear in unilateral aural atresia.
- Although the uninvolved ear was normal in all the cases from our series, a comprehensive assessment of hearing is necessary. In the rare case of bilateral hearing impairment, early intervention is necessary.

References

1. Jahrsdoerfer RA. Congenital atresia of the ear. *Laryngoscope.* 1978;88 (Suppl 13):1-48.
2. Declau F, Cremers C, Van de Heyning P. Diagnosis and management strategies in congenital atresia of the external auditory canal. Study Group on Otological Malformations and Hearing Impairment. *Br J Audiol.* 1999;33:313-327.
3. Okajima H, Takeichi Y, Umeda K, Baba S. Clinical analysis of 592 patients with microtia. *Acta Otolaryngol Suppl.* 1996;525:18-24.
4. Korres SG, Balatsouras DG, Economou C, Ferekidis E, Kandiloros D, Adamopoulos G. Effect of the number of averaged responses in transient evoked otoacoustic emissions on the results of neonatal hearing screening. *Audiology.* 2000;39:293-299.
5. Brent B. Technical advances in ear reconstruction with autogenous rib cartilage grafts: personal experience with 1200 cases. *Plast Reconstr Surg.* 1999;104:319-338.
6. Mayer TE, Brueckmann H, Siegert R, Witt A, Weerda H. High-resolution CT of the temporal bone in dysplasia of the auricle and external auditory canal. *AJNR Am J Neuroradiol.* 1997;18:53-65.
7. Coccaro PJ, Becker MH, Converse JM. Clinical and radiographic variations in hemifacial microsomia. *Birth Defects Orig Artic Ser.* 1975;11:314-324.
8. Altmann F. Congenital atresia of the ear in man and animals. *Ann Otol Rhinol Laryngol.* 1955;64:824-858.
9. Watkin PM. Neonatal screening for hearing impairment. *Semin Neonatol.* 2001;6:501-509.
10. Korres SG, Balatsouras DG, Gkoritsa E, Kandiloros D, Korres GS, Ferekidis E. The effect of very low birth weight on otoacoustic emissions. *B-ENT.* 2007;3:15-20.
11. Bray P, Kemp D. An advanced cochlear echo technique suitable for infant screening. *Br J Audiol.* 1987; 21:191-204.
12. Suutarla S, Rautio J, Ritvanen A, Ala-Mello S, Jero J, Klockars T. Microtia in Finland: comparison of characteristics in different populations. *Int J Pediatr Otorhinolaryngol.* 2007;71: 1211-1217.
13. McKinnon BJ, Jahrsdoerfer RA. Congenital auricular atresia: update on options for intervention and timing of repair. *Otolaryngol Clin North Am.* 2002;35:877-890.

Dimitrios Balatsouras, M.D.
23 Achaion Street – Ag. Paraskevi
GR-15343 Athens, Greece
Tel.: +30 210 600 4683,
Fax: +30 210 4592 671
E-mail: balats@vodafone.net.gr