

The audiological characteristics of infant auditory neuropathy patients without otoacoustic emission: A retrospective clinical study

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Abstract

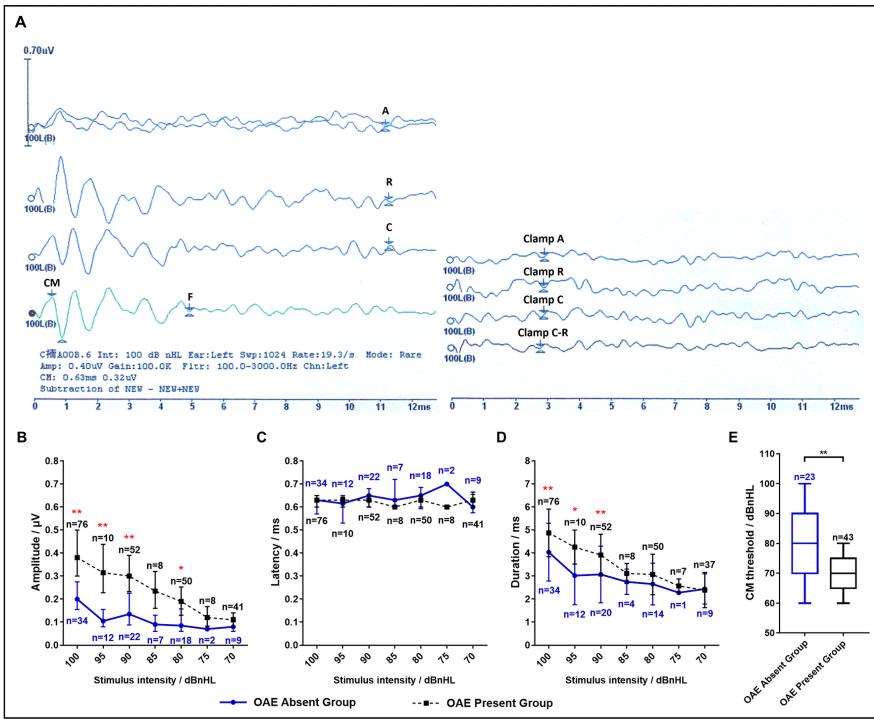
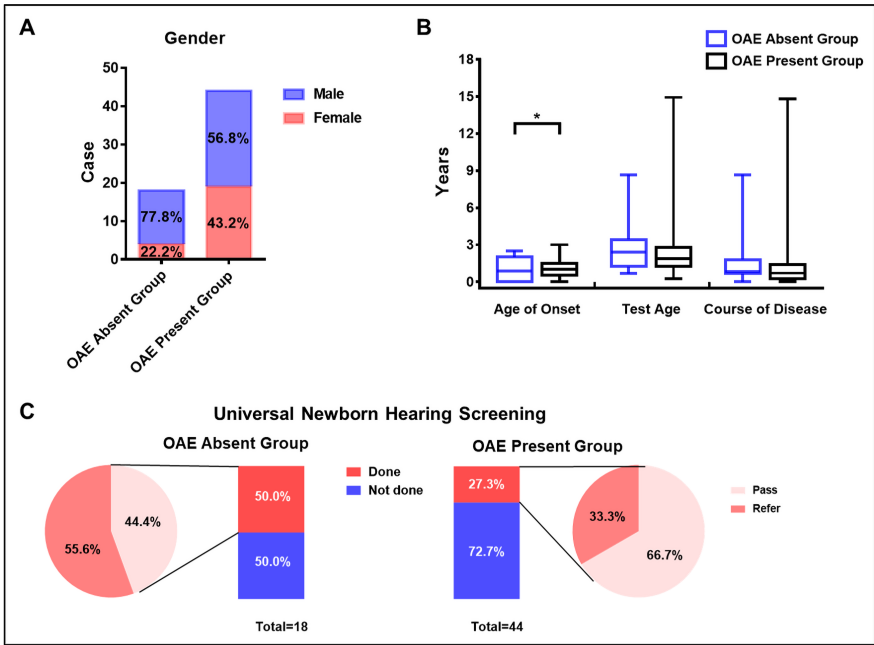
Objective: To explore the audiological characteristics of infant auditory neuropathy (AN) patients with cochlear microphonic (CM) recorded but no otoacoustic emission (OAE) response and clinically reduce the rate of missed diagnosis of AN. **Design:** Retrospective clinical study of medical data from 2003 to 2020. **Setting:** Otolaryngology head and neck surgery clinical hearing center. **Participants:** Eighteen infant AN patients with CM present and distortion product otoacoustic emission (DPOAE) absent in both ears were OAE absent group. Forty-four infant AN patients with CM and DPOAE present in both ears were OAE present group. **Main outcome measures:** Audiological characteristics. **Results:** 1. The age of onset in OAE absent group was 0.9 (0.02) years old, which was less than 1.11 (1.63) years old in OAE present group ($P=0.041$). 2. The CM threshold of OAE absent group was 80 (10) dB nHL, which was significantly higher ($P<0.001$) than OAE present group. CM amplitude were smaller ($P<0.05$), and CM duration were shorter ($P<0.05$) in OAE absent group. 3. The thresholds of auditory steady-state response (ASSR) at 0.5, 1, 2 and 4 kHz were 94 (10), 94 (10), 87 (20) and 81 (10) dB HL cg respectively in OAE absent group, which were higher than those in OAE present group ($P<0.01$). **Conclusions:** Infant AN patients with CM present and OAE absent showed earlier onset, worse hearing level and worse CM performance. The influencing factors and value of CM in AN patients still need to be explored in the future.

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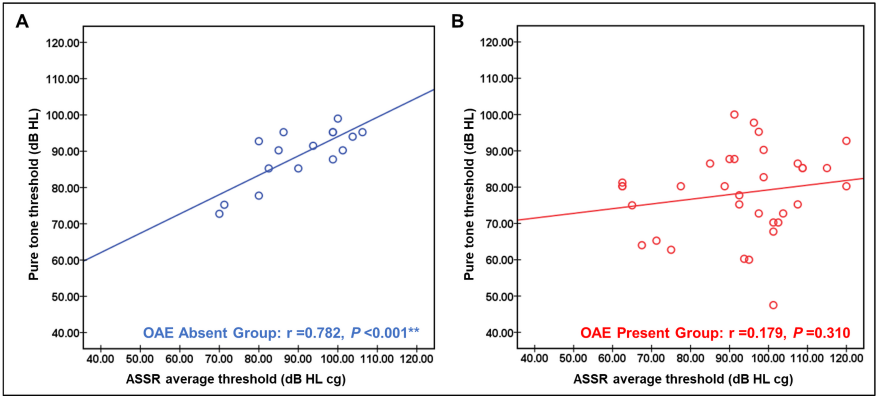
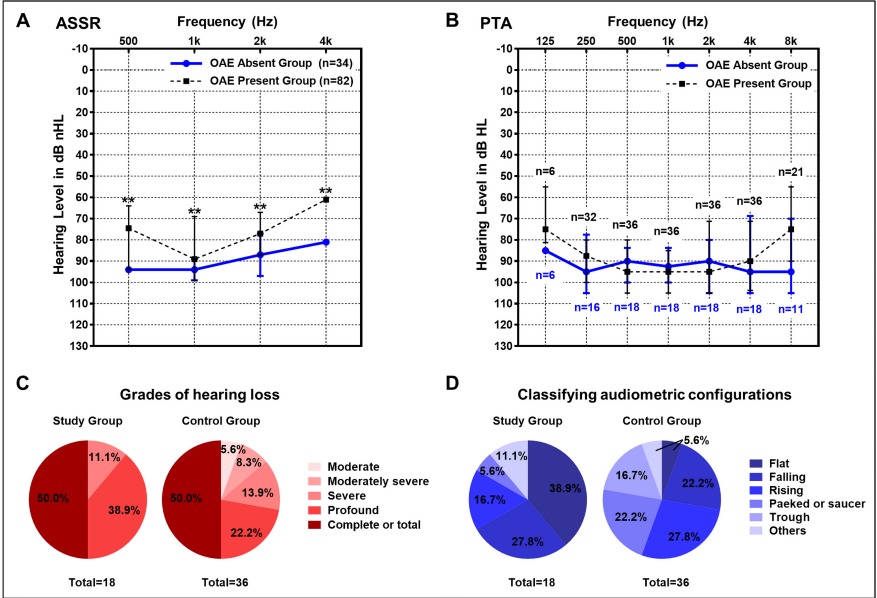


Table 1. Audiological Characteristics of Patients.

Characteristic	OAE absent Group (n=36)	OAE present Group (n=88)	P Value
Pure tone audiometry/behavioral audiometry			
Grades of hearing loss - no./total no. (%)			
Mild	0	0	0.525
Moderate	0	2/36 (5.6)	
Moderately severe	0	3/36 (8.3)	
Severe	2/18 (11.1)	5/36 (13.9)	
Profound	7/18 (38.9)	8/36 (22.2)	
Complete or total	9/18 (50.0)	18/36 (50.0)	
Classifying audiometric configurations - no./total no. (%)			
Flat	7/18 (38.9)	2/36 (5.6)	0.013*
Falling	5/18 (27.8)	8/36 (22.2)	
Rising	3/18 (16.7)	10/36 (27.8)	
Peaked or saucer	1/18 (5.6)	8/36 (22.2)	
Trough	0	6/36 (16.7)	
Others	2/18 (11.1)	2/36 (5.6)	
0.5-4 kHz PTA - M (P ₂₅ , P ₇₅) dB HL	96.25 (81.88, 100.31)	94.38 (75.63, 102.19)	0.993
ABR - no./total no. (%)			
Absent	34/36 (94.4)	83/88 (94.3)	0.080
Abnormal			
Only wave I at 100 dB nHL	0	1/88 (1.1)	
Wave V latency delay at 100 dB nHL	2/36 (5.6)	0	
The threshold increased significantly	0	4/88 (4.5)	
CM			
Amplitude at 100 dB nHL - M (P ₂₅ , P ₇₅) μV	0.20 (0.16, 0.28)	0.38 (0.30, 0.50)	<0.001**
Latency at 100 dB nHL - M (P ₂₅ , P ₇₅) ms	0.63 (0.57, 0.65)	0.63 (0.6, 0.65)	0.823
Duration at 100 dB nHL - Mean ± SD ms	4.04 ± 1.25	4.88 ± 1.03	0.001**
Threshold - dB nHL	80.00 (70.00, 90.00)	70.00 (65.00, 75.00)	<0.001**
ASSR threshold - M (P ₂₅ , P ₇₅) dB HL cg			
500Hz	94.00 (84.00, 94.00)	74.50 (64.00, 94.00)	<0.001**
1000Hz	94.00 (89.00, 99.00)	89.00 (69.00, 99.00)	0.003**
2000Hz	87.00 (77.00, 97.00)	77.00 (67.00, 87.00)	<0.001**
4000Hz	81.00 (71.00, 81.00)	61.00 (61.00, 71.00)	<0.001**
40Hz-AERP threshold - M (P ₂₅ , P ₇₅) dB nHL	100.00 (100.00, 110.00)	100.00 (100.00, 110.00)	0.399

no.: number; n: number of ears; M (P₂₅, P₇₅): Median (upper quartile, lower quartile); ABR: auditory brainstem response; CM: cochlear microphonic; ASSR: auditory steady-state response; 40Hz-AERP: 40Hz-auditory event related potential.

*: $P < 0.05$; **: $P < 0.01$.

Table 2. Follow-up data of three cases in the OAE absent group

Characteristic		Case 1		Case 2		Case 3	
Gender		M		M		M	
Age of onset - yr		2.00		1.00		1.50	
Newborns high risk factors		None		None		Unknow	
Universal Newborn Hearing Screening		Not done		Pass		Pass	
Concomitant symptoms		None		None		None	
Test age - yr		3.50	4.00	1.42	3.00	2.17	5.50
Course of disease - yr		1.50	2.00	0.42	2.00	0.67	4.00
PTA - dB HL	L	80.00	78.75 ↓	NA	97.50	NA	123.75
	R	86.25	91.25 ↑	NA	110.00	NA	108.75
Classifying audiometric configurations	L	Rising	Rising	NA	Rising	NA	Flat
	R	Rising	Rising	NA	Rising	NA	Flat
Tympanogram	L/R	A/A	NA	A/A	A/C	A/A	C/C
Acoustic stapedial reflex		NR	NA	NR	NR	NR	NA
DPOAE	L	NR	NR	NR	NR	NR	NR
	R	NR	NR	NR	NR	NR	NR
ABR	L	NR	NR	V=6.35ms	NR	NR	NR
	R	NR	NR	V=6.00ms	NR	NR	NR
CM (100 dB nHL)	Amplitude (L) - μV	0.46	0.32 ↓	0.14	0.08 ↓	0.13	NR ↓
	Latency (L) - ms	0.70	0.60 ↓	0.72	0.82 ↑	0.63	NR ↓
	Duration (L) - ms	6.65	4.83 ↓	2.30	2.17 ↓	3.63	NR ↓
	Amplitude (R) - μV	0.38	0.27 ↓	0.29	NR ↓	0.19	NR ↓
	Latency (R) - ms	0.57	0.57 -	0.65	NR ↓	0.60	NR ↓
	Duration (R) - ms	6.53	3.17 ↓	3.42	NR ↓	3.98	NR ↓
CM threshold - dB nHL	L	60	75 ↑	70	100 ↑	NA(<100)	100 ↑
	R	70	75 ↑	75	NR ↑	NA(<100)	NR ↑
ASSR - dB nHL	L	92.75	NA	92.75	NR ↑	99.00	NA
	R	90.25	NA	84.00	NR ↑	85.88	NA
40Hz-AERP - dB nHL	L	110	NA	120	NA	110	NA
	R	110	NA	120	NA	110	NA

yr: years; PTA: pure tone audiometry; DPOAE: distortion product otoacoustic emission; ABR: auditory brainstem response; CM: cochlear microphonic; ASSR: auditory steady-state response; 40Hz-AERP: 40Hz-auditory event related potential.
 NA: not available; NR: no reaction.