

Jadranka Handzic M.D., Ph.D.,
Professor of Otolaryngology, Audiology and Vestibulology
Department for Otolaryngology and Audiology
University Hospital Center “Rebro”
Kispaticeva 12
Croatia

HEARING LOSS IN UNILATERAL CLEFT LIP AND PALATE



EMBRYOLOGY

-Cleft lip and/or palate- one of the most frequent structural defect (1st branchial arch) of the head and neck

- fail of fusion of left and right mesoderm plate in the middle line

primary palate origin at 6th weeks of gestation

-secondary palate - 8-10th hard palate

- 10-12th soft palate



ETIOLOGY-INCIDENCE

- **Etiology-** multifactorial
- **-Incidence** of isolated cleft lip and palate:
 - 1 in 750 live births
 - left unilateral cleft lip and palate occurs twice as many males than females



PATHOANATOMICAL CRANIOFACIAL FEATURES-HIGH RISK OF LONG-TERM NEGATIVE MIDDLE EAR PRESSURE

Cranial base- changes of the length and angulations of anterior cranial base, smaller sphenopalatine angle

Eustachian tube - hypoplastic and hypoelastic cartilaginous part, inadequate clearance and ventilation of the middle ear/development connection with growing of the middle face

Maxilla – retrognathia, retarded middle face

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Pharyngeal latero-lateral distance- increase, cranio-caudal distance-decrease, pathologic movement of the lateral wall, Passavant or circular ridge during deglutition

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Middle ear cavity - “small mastoid and middle ear cavity”-

Cleft palate -short and high positioned-delay in descending

Cleft palate muscles-hypoplastic and malpositioned tensor and levator veli palatini, neuromuscular delay of maturity of the pharyngeal muscles

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MORPHOLOGICAL CHANGES IN UNILATERAL CLEFT LIP AND PALATE



- retrognathic and retarded growth of maxilla
- influence on the growth of the Eustachian tube region
- lower position of auricula,
- retarded growth of mandibula
- collapse of the top of the nose and alar rotation,
- relative hypertelorism

CLEFT LIP AND PALATE CHILDREN-UNIVERSALITY OF THE OTITIS MEDIA WITH EFFUSION

-long term negative middle ear pressure leads to edema of the middle ear mucosa and **accumulation of the fluid in the middle ear;**

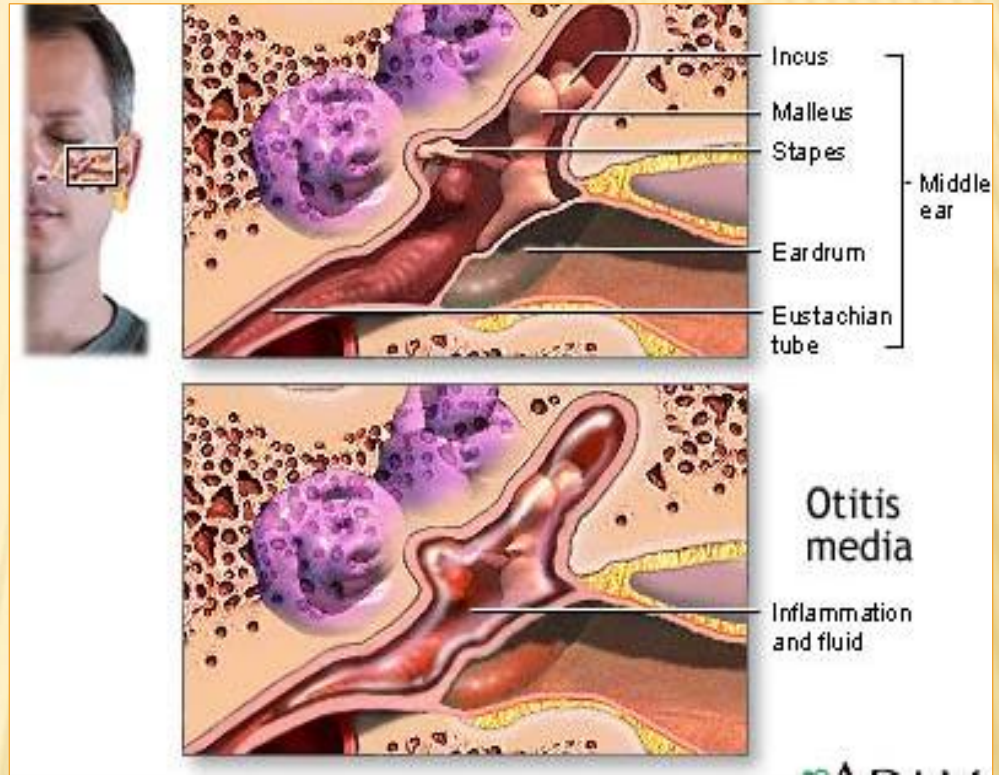
Otitis media with effusion (OME)- the most common disease in childhood

- the most frequent cause of communication disorders

OME-usually finding in the left lip and palate children until 3yr.

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OTITIS MEDIA WITH EFFUSION

Otoscopic finding

Accumulation of the middle ear fluid

CONDUCTIVE HEARING LOSS-EFFECT ON CENTRAL AUDITORY PROCESSING -SPEECH DISCRIMINATION

- ✘ **-distortion of the sound reaching cochlea**-interaural delay in sound reaching the brain
 - ✘ **-disturbance of binaural central auditory processing** ,disturbed inhibitory process on neural synapses
 - disturbance in neural encoding and tonotopic organization of primary auditory cortex, making short and long-term **auditory working memory**
 - ✘ **-negative influence** on development **of speech and language** acquisition, phonemic awareness-speech discrimination, cognition, fine motoric function, long term acoustic memory, balance, sound segregation in background noise-
 - ✘ **Otitis media with effusion leads to central auditory processing deficit even after the restoration of peripheral function and normalization of the hearing threshold**
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UNILATERAL CLEFT LIP AND PALATE (UCLP) VS. BILATERAL CLEFT LIP AND PALATE (BCLP) – COMPARATIVE STUDY OF HEARING LOSS

- 1) UCLP - **higher incidence of hearing loss ears (89%)** at age 1-3yr. than BCLP (62%)
- 2) UCLP - **lower incidence of ears with normal hearing threshold** than BCLP
- 2) UCLP - **higher incidence of ears with moderate and severe hearing loss** than BCLP
- 3) UCLP - **moderate hearing loss- slower** improvement of hearing threshold than BCLP

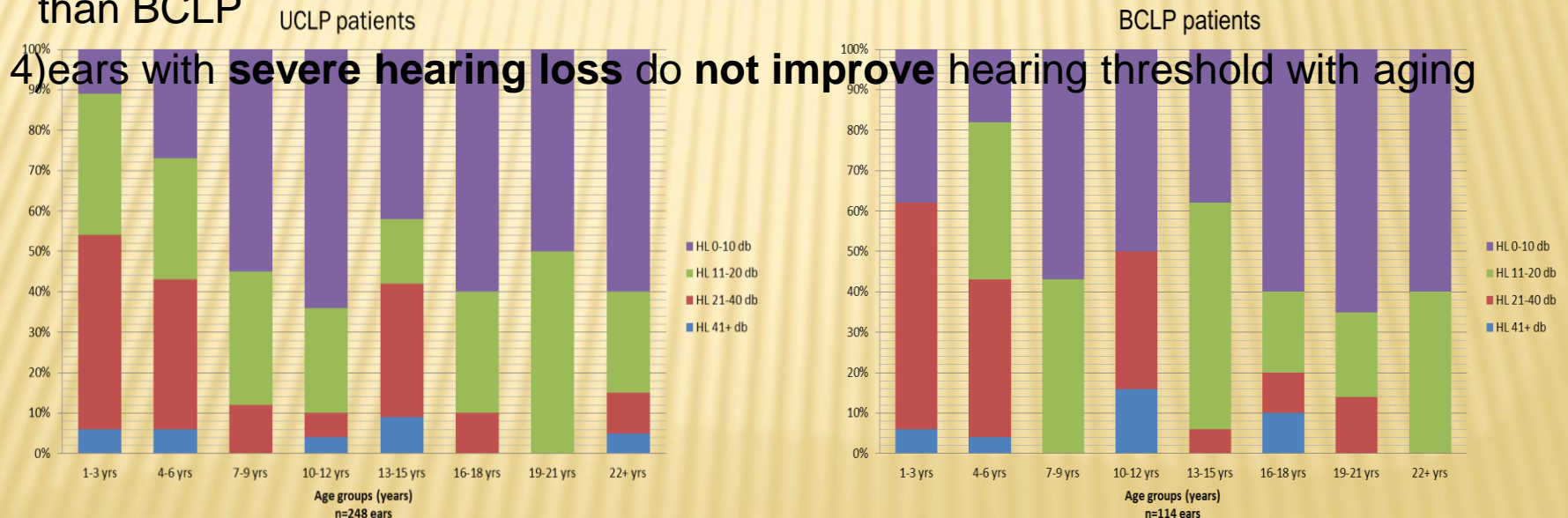


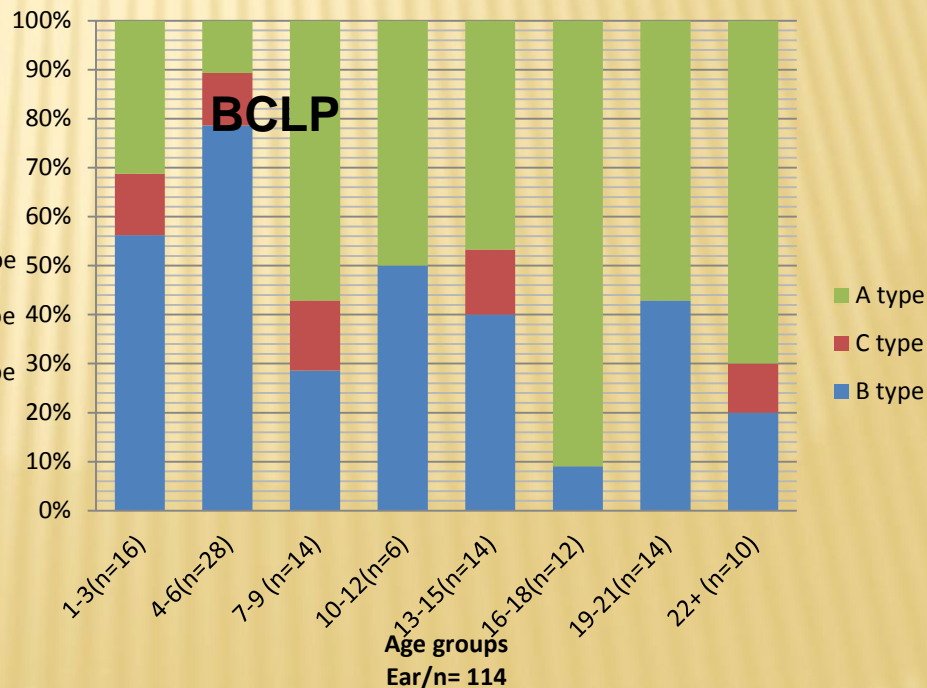
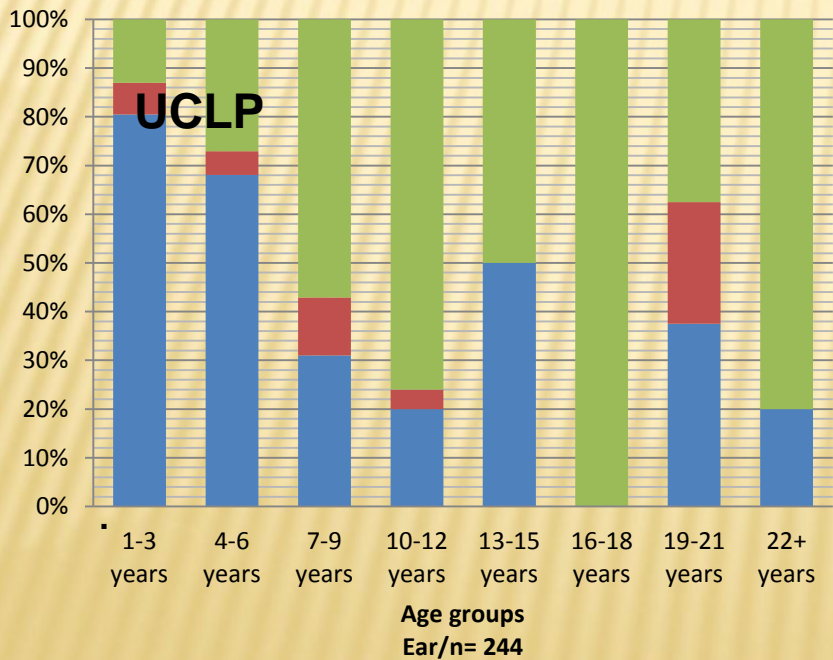
Fig. 3. Proportions of ears with mild, moderate and severe hearing loss (stacked) according to age in UCLP group. Significant negative correlation with age was found for moderate hearing loss.

Fig. 2. Proportions of ears with mild, moderate and severe hearing loss (stacked) according to age in BCLP patients. Significant negative correlation with age was found for moderate (21 -40 dB) hearing loss.

TYMPANOMETRIC FINDINGS IN UCLP VS. BCLP EARS-COMPARATIVE STUDY

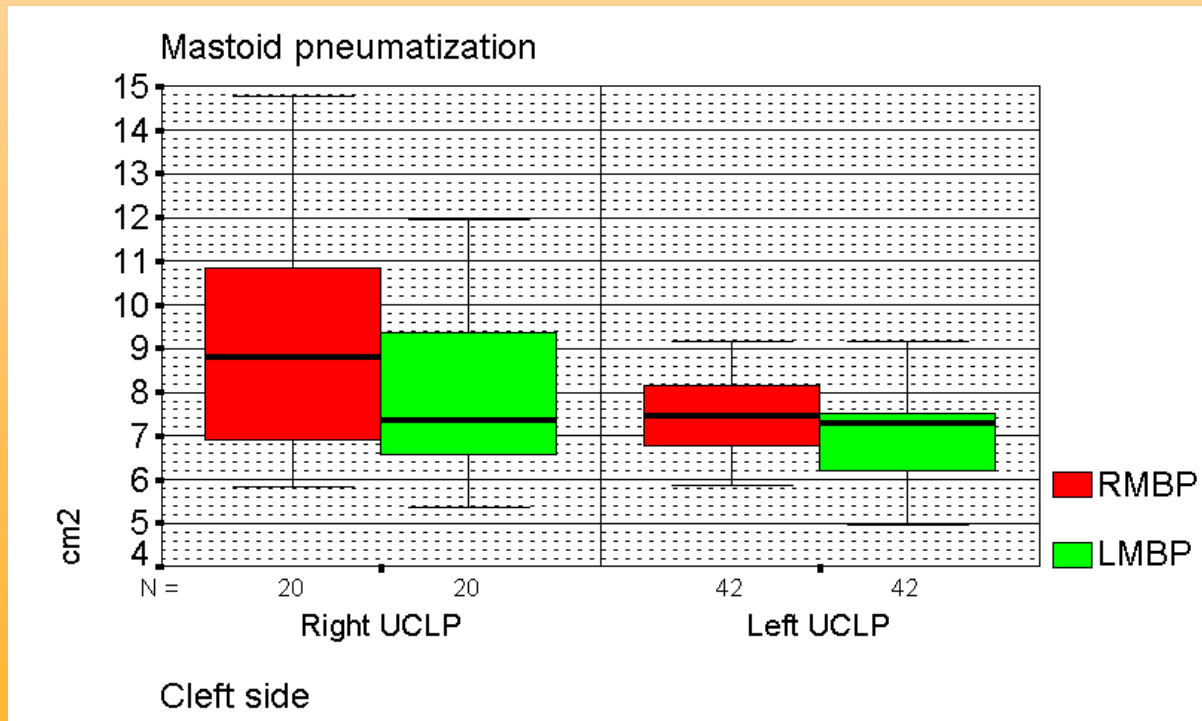
UCLP- B type – frequency higher at age 1-3yr than in BCLP and ICP

- decreased faster than in the BCLP and ICP
- UCLP decrease of B type from age 7-9yr, BCLP 4-6yr, ICP from 15yr.



MATOID BONE PNEUMATIZATION IN UNILATERAL CLEFT LIP AND PALATE OF LEFT VS. RIGHT SIDE

UCLP-right and UCLP-left - mastoid bone pneumatization **of left ears** - smaller than mastoid bone pneumatization of right ears



AIM OF THE STUDY AND HYPOTHESIS



- × **Different pathoanatomical and developmental characteristics of anatomical structures of cleft vs. non cleft side have different mode of pathophysiological development of middle ear disease**



METHOD

Study included 101 children: median age 6.0 yr.,

- 68 males

-33 females

- Palatoplasty and cheiloplasty performed previously according to standard protocol

Hearing loss (tonal audiometry and tympanometry) analyzed according to age subgroups :

1-3 years,4-7years,8-12 years

AUDIOLOGIC TESTS-TYMPANOMETRY-TONAL AUDIOMETRY



- × **Tympanometry-** types according to **Jerger; A,C,B**
- × **Tonal audiometry;**
- × Tested audiometric frequencies ;
250Hz,500Hz,1000Hz,2000Hz,4000Hz
- × Normal hearing threshold: 0-10dB
- × -mid hearing loss; 11-20dB
- × -moderate hearing loss;21-40dB
- × -severe hearing loss >40dB
- × Results described and compared
- × -median (Md) hearing level for tested frequencies
- × - average hearing level (AHL) across speech frequencies

Age (years) UCLP	N (76)	Ear side	Hearing level (dB)						Kruskal-Wallis test (p value)
			AHL	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	
			Md	Md	Md	Md	Md	Md	
			(Min-Max)	(Min-Max)	(Min-Max)	(Min-Max)	(Min-Max)	(Min-Max)	
1-3 yr.	30	RIGHT ears	p=0.683 35.0 (16-41)	p=0.398 30.0 (20-40)	p=0.043 40.0 (20-50)	p=0.866 32.5 (15-50)	p=0.906 30.0 (15-45)	p=0.612 30.0 (10-50)	p=0.057 p=0.601
		LEFT ears	33.0 (20-42)	27.5 (20-45)	30.0 (15-40)	32.5 (15-45)	32.5 (15-50)	32.5 (15-50)	
4-7yr.	57	RIGHT ears	p=0.039 21.0 (10-47)	p=0.387 25.0 (10-45)	p=0.052 25.0 (10-55)	p=0.054 20.0 (10-45)	p=0.410 20.0 (10-45)	p=0.003 20.0 (10-55)	p=0.001 p=0.003
		LEFT ears	25.0 (10-47)	25.0 (10-50)	30.0 (10-45)	30.0 (10-50)	25.0 (10-50)	25.0 (10-50)	
8-12yr.	17	RIGHT ears	p=0.906 20.0 (10-41)	p=0.790 20.0 (10-45)	p=0.650 25.0 (10-35)	p=0.979 20.0 (10-50)	p=0.875 20.0 (10-30)	p=0.382 20.0 (10-55)	p=0.294 p=0.028
		LEFT ears	18.0 (10-47)	20.0 (10-45)	20.0 (10-45)	20.0 (10-55)	15.0 (10-40)	20.0 (10-55)	
>12yr.	12	RIGHT ears	p=0.110 16.0 (10-42)	p=0.787 12.0 (10-35)	p=0.866 15.0 (10-50)	p=0.590 17.5 (10-40)	p=0.043 10.0 (10-40)	p=0.052 10.0 (10-45)	p=0.269 p=0.972
		LEFT ears	18.0 (10-44)	17.5 (10-40)	15.0 (10-40)	17.5 (10-50)	17.0 (10-50)	17.5 (10-60)	



RESULTS

- × **Age group 1-3yr.**
- × - non cleft side - highest severity of hearing loss (Md) 500Hz severity of hearing loss than lefts
- × **Age group 4-7yr. ;**
- × AHL higher on cleft side
- × - cleft side ears -higher severity of hearing loss for mid register (Md)(500Hz,1000Hz,4000Hz)
- × -non cleft side-higher severity of hearing loss for low register (250Hz and 500Hz)

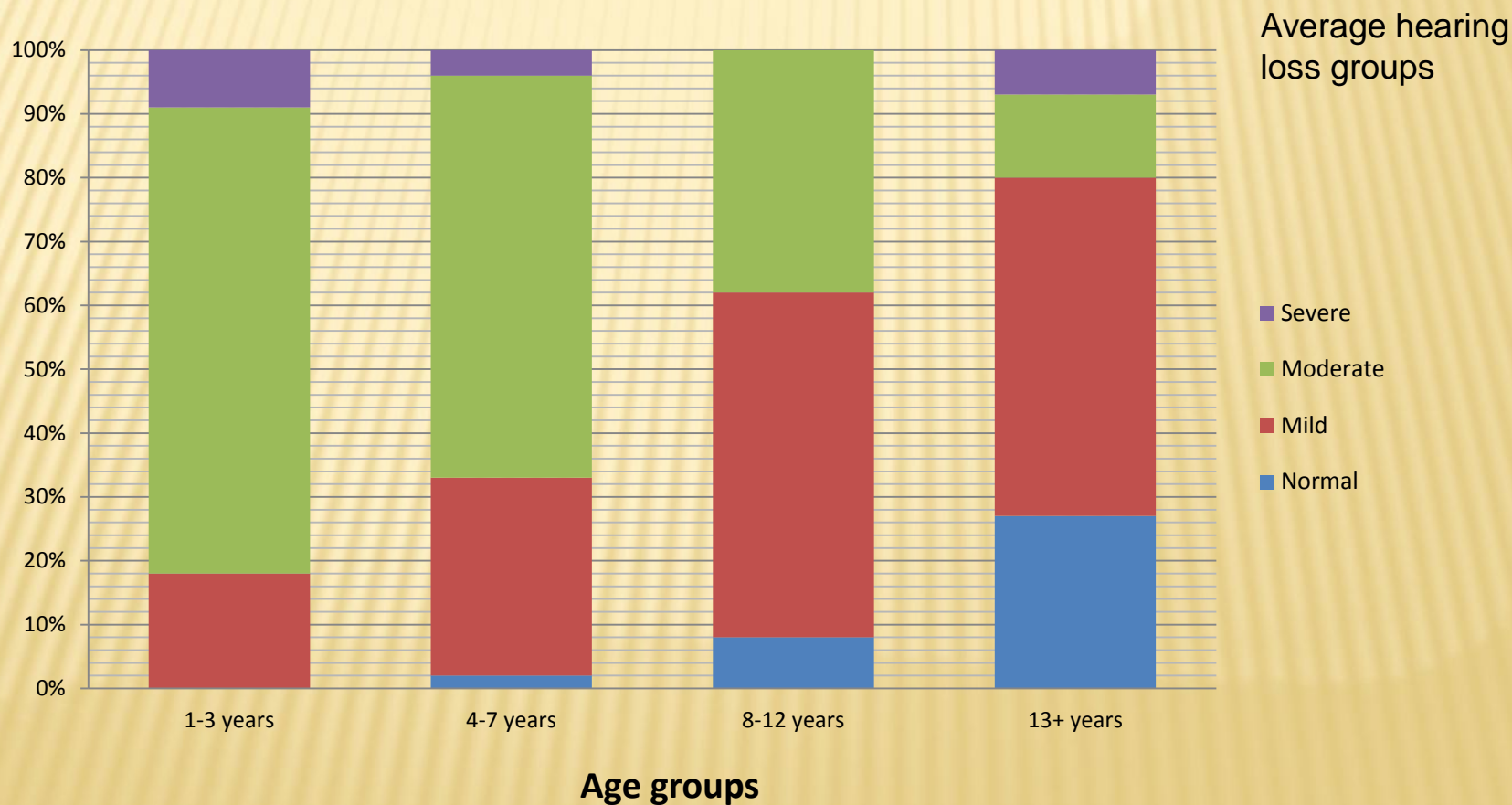
RESULTS



- × **Age group 8-12 yr;**
 - × hearing loss cleft vs.non cleft side showed no significant difference
 - × Cleft side ears- higher hearing loss on frequencies 500Hz,1000Hz then on others
- Age group >12 yr;**
- ear side dependence in severity of hearing loss showed frequencies of 2000Hz and 4000Hz-cleft side showed higher severity

UCLP- LEFT EAR (CLEFT SIDE)

PROPORTION OF EARS WITH MILD, MODERATE AND SEVERE HEARING LOSS (AVERAGE) ACCORDING TO AGE SUBGROUPS



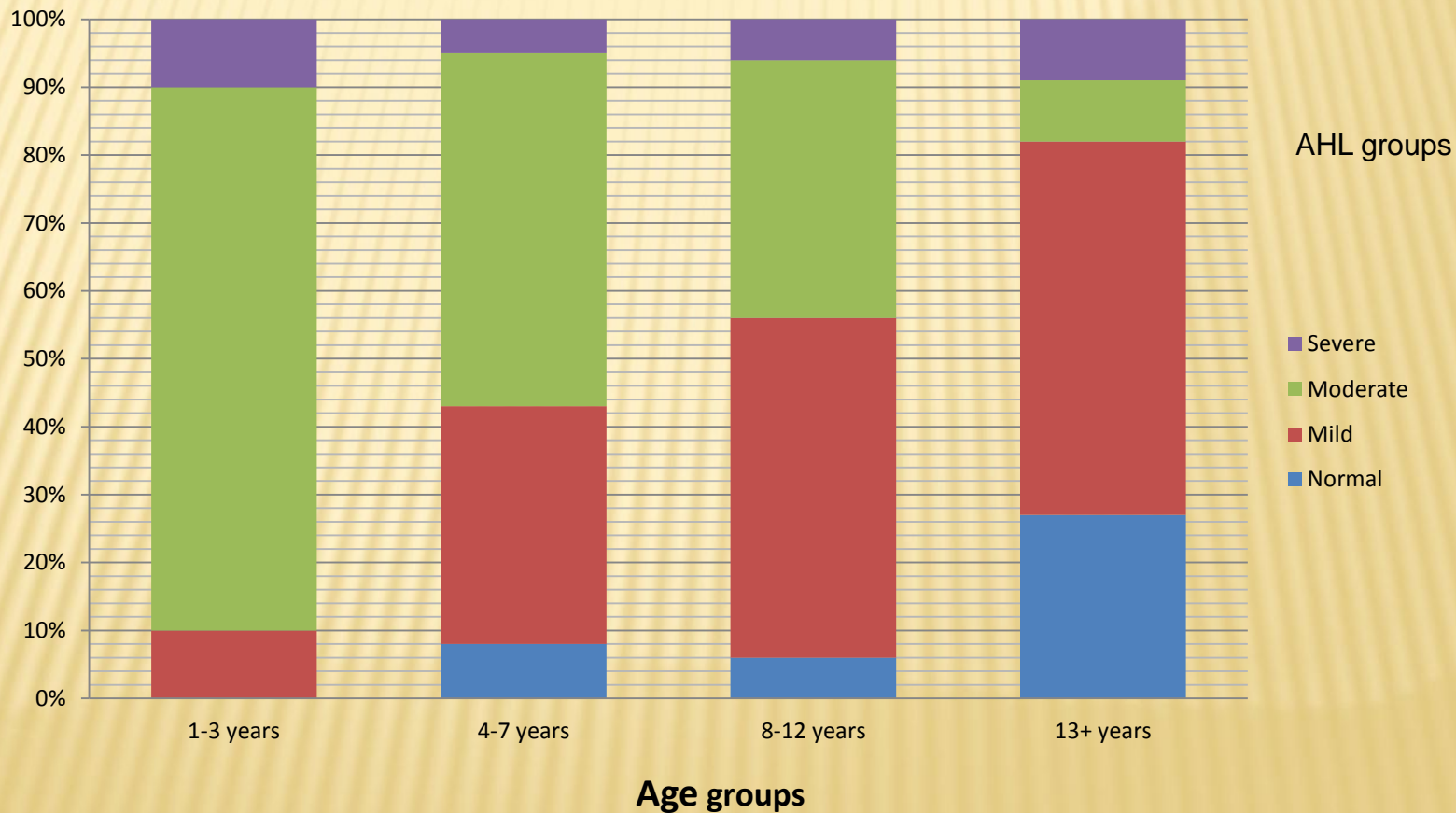
RESULTS-CLEFT SIDE EARS



- ✘
- ✘ **-age group 1-3yr.** -80% of ears have moderate hearing loss (21-40dB)
 - incidence of ears with moderate hearing loss significantly decrease with aging**
- ✘ **-no increase of incidence of ears with normal hearing level**
- ✘ **with aging**
 - intergroup comparison of hearing loss ears showed significant difference for moderate category**

UCLP- RIGHT EAR (NON-CLEFT SIDE)

PROPORTION OF EARS WITH HEARING LOSS (AVERAGE HL)- MILD, MODERATE, SEVERE ACCORDING TO AGE SUBGROUPS



RESULTS-NON CLEFT EARS



- × **Non cleft side ears ;**
- × Increased rate of ears with normal hearing threshold with aging
- × Intergroup comparison showed significant difference for **mid** and **moderate** category of hearing loss ears
- × Intragroup comparison difference showed age group 4-7yr

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IMPROVEMENT OF HEARING THRESHOLD WITH AGING

TABLE 2
ABSOLUTE DIFFERENCE IN AHL AND MHL (DB) BETWEEN NEIGHBORING AGE GROUPS

Cleft type Age groups compared	Right ear						Left ear					
	AHL.R	AHL.L	250 Hz	500 Hz	1000 Hz	2000 Hz	4000 Hz	250 Hz	500 Hz	1000 Hz	2000 Hz	4000 Hz
UCLP												
1-3 vs. 4-7	-14*	-8	-5	-20*	-10*	-10*	-5*	0	-5	-5	-10	-5
4-7 vs. 8-12	-3	-7**	-5	0	0	-5	0	-5*	-5**	-5*	-5**	-5*
8-12 vs. 13+	-2	-2	0	-5	-5	-5	-10*	-5	-5	-10	-5	-5
UCLP (R)												
1-3 vs. 4-7	-	-	-	-	-	-	-	-	-	-	-	-
4-7 vs. 8-12	-6	-3	-5	0	-5	-8	-8	-8*	-5	-2	-5	0
8-12 vs. 13+	-	-	-	-	-	-	-	-	-	-	-	-
UCLP (L)												
1-3 vs. 4-7	-14*	-8	-5	-15**	-13*	-10**	-10*	-3	0	-3	-8	-8
4-7 vs. 8-12	-1	-9*	-5	-3	0	-3	3	-5	-10*	-10*	-12	-7*
8-12 vs. 13+	-4	1	0	-8	-5	-8	-13	-5	-5	-5	2	-3

* statistically significant at $p \leq 0.05$; ** $p \leq 0.01$

AHL.R=average hearing level - right ears; AHL.L=average hearing level - left ears; UCLP=unilateral cleft lip and palate; UCLP (R)=unilateral cleft lip and palate (right side); UCLP (L)=unilateral cleft lip and palate (left side).

CONCLUSION

- ✘ **Cleft side ears :**
- ✘ **showed different pathophysiology of OME than none cleft side and difference in severity of hearing loss for restrictive frequencies in dependence with ear side and age**
- ✘ **characteristics of hearing loss and its improvement with aging determine speech and language habilitation and prevention of central auditory disorders**

WARM GREETINGS FROM ZAGREB, CROATIA

