Temporal bone imaging in osteogenesis imperfecta patients with hearing loss

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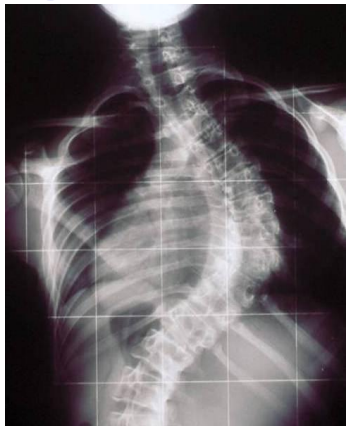
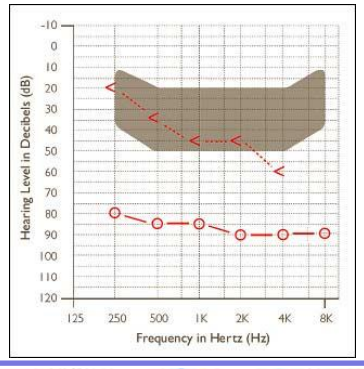
I. Introduction

Osteogenesis Imperfecta (OI)

- ‘Brittle bone disease’
- Connective tissue disease (1/20.000)

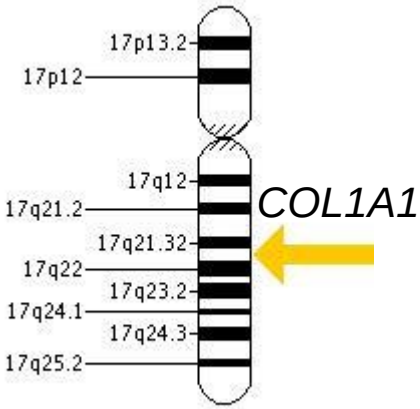
Phenotype

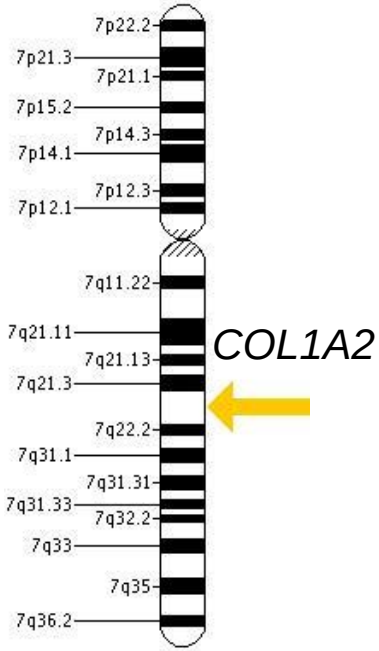




Genotype

Autosomal dominant mutation (90%)



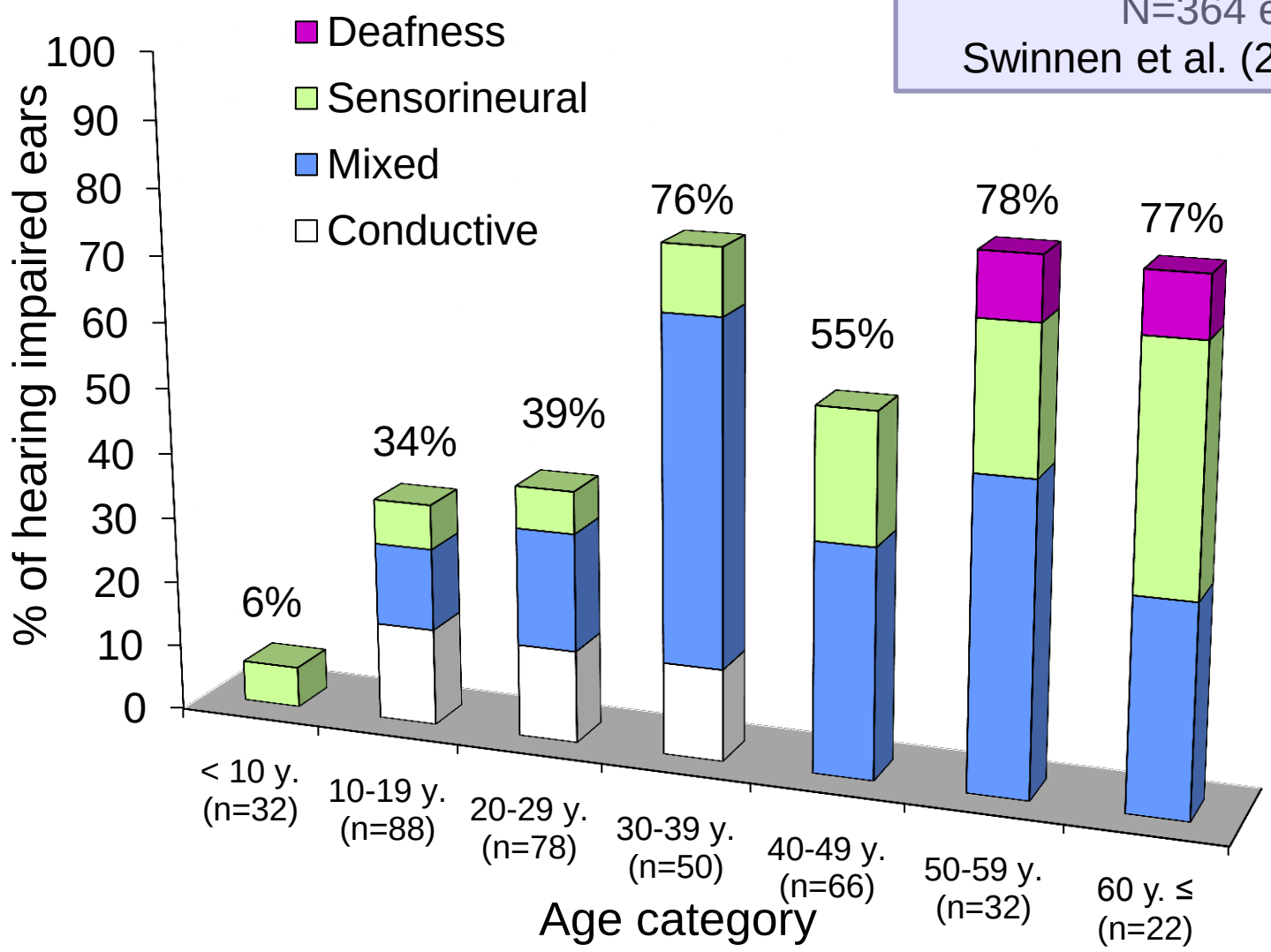


Impaired synthesis of type I collagen

I. Introduction

OI – Hearing loss: prevalence and type

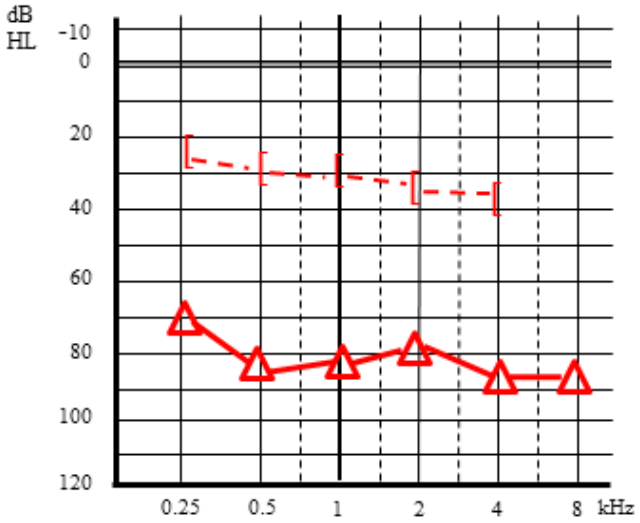
N=364 ears
Swinnen et al. (2012)



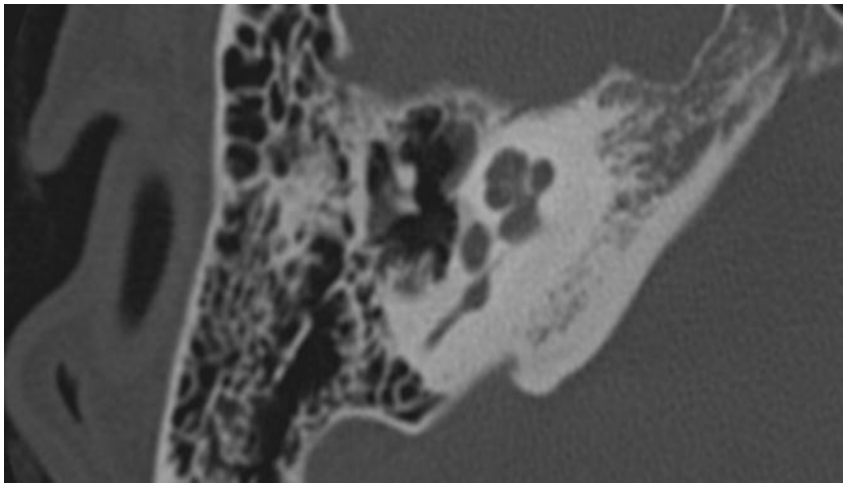
I. Introduction
Research aims

Association between temporal bone imaging findings and audiometric results?

Audiogram



CT temporal bone



II. Methods
Patients and materials

- 17 patients
  - Age: 9-67 y.
  - *COL1A1* or *COL1A2* mutation

- Retrospective study
  - Audiograms
  - CT images temporal bone (17 patients; 33 ears)

III. Results
Audiometric profiles

- Hearing loss in 31/33 ears
- Type of hearing loss

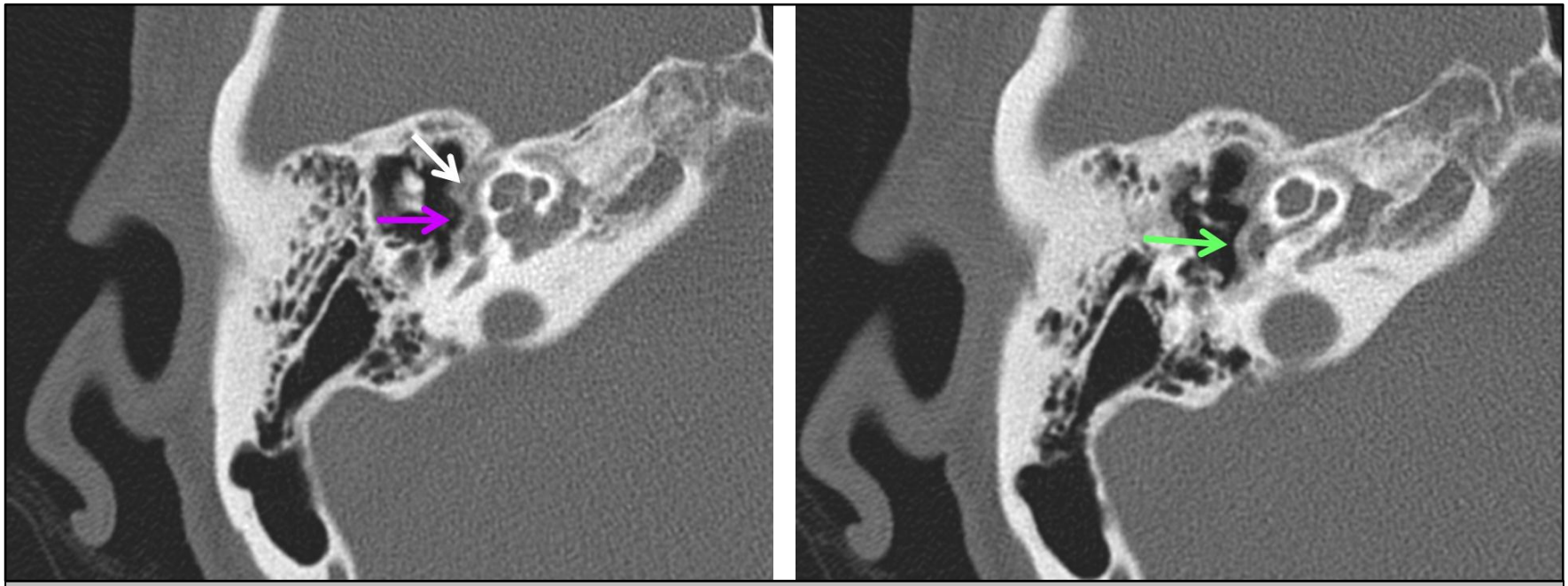
	No. of ears
Normal hearing	2
Conductive hearing loss	4
Mixed hearing loss	20
Sensorineural hearing loss	2
Deafness	5

- Hearing loss severity: mild to profound

III. Results
Radiological evaluation: CT (1)

Fenestral hypodensities in 26/33 ears (79%)

FAF	25/33	76%
Oval window	23/33	70%
Round window	20/33	61%



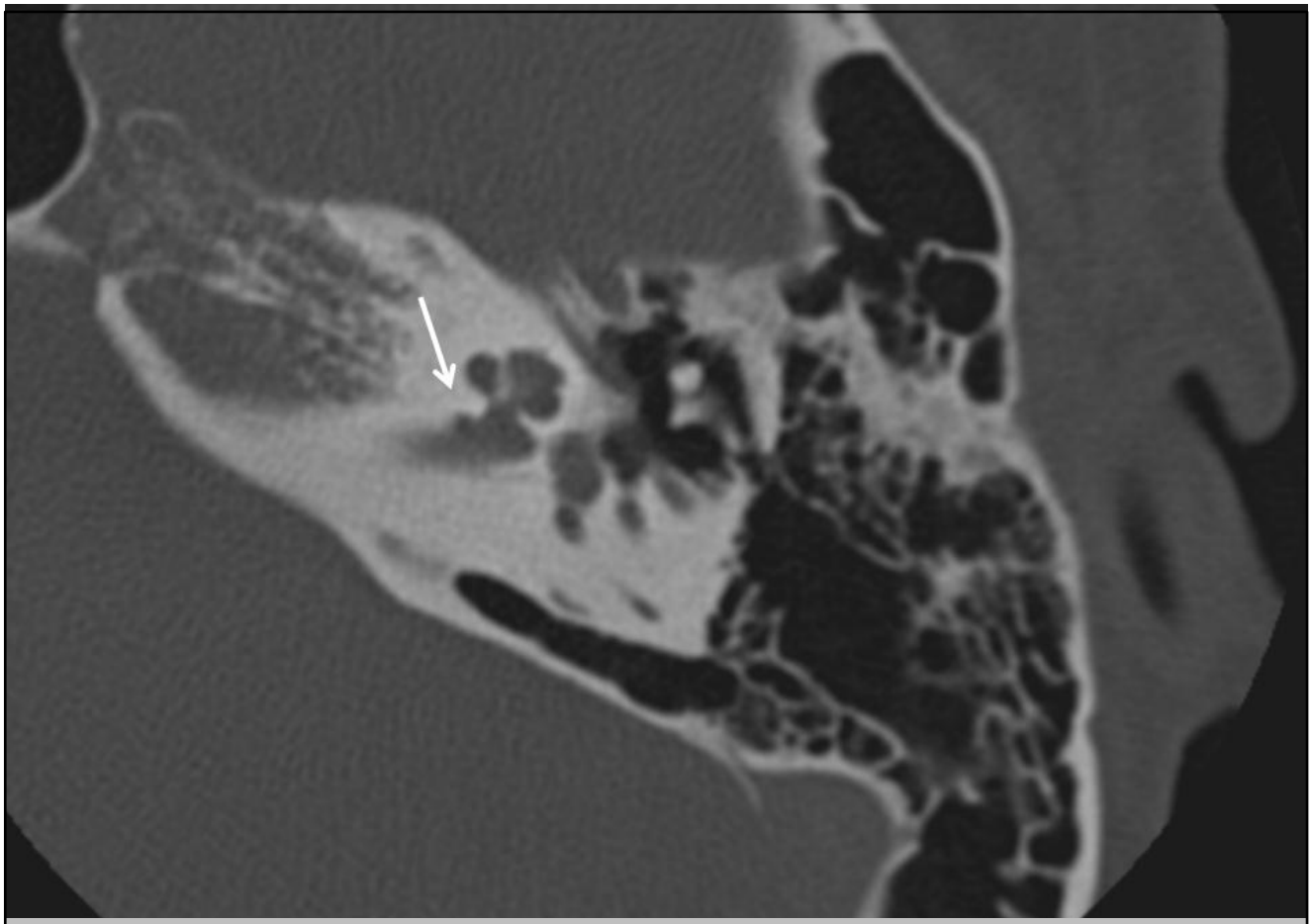
Axial CT images through right temporal bone (mixed hearing loss)

III. Results
Radiological evaluation CT (2)

Retrofenestral hypodensities in 20/33 ears (61%)

Cochlear turns	16/33	49%
Hypodensity antero-inferior to internal auditory canal	16/33	49%
Double ring sign	11/33	33%
Facial nerve canal	10/33	30%
Semicircular canals	6/33	18%

Radiological evaluation CT: retrofenestral hypodensities

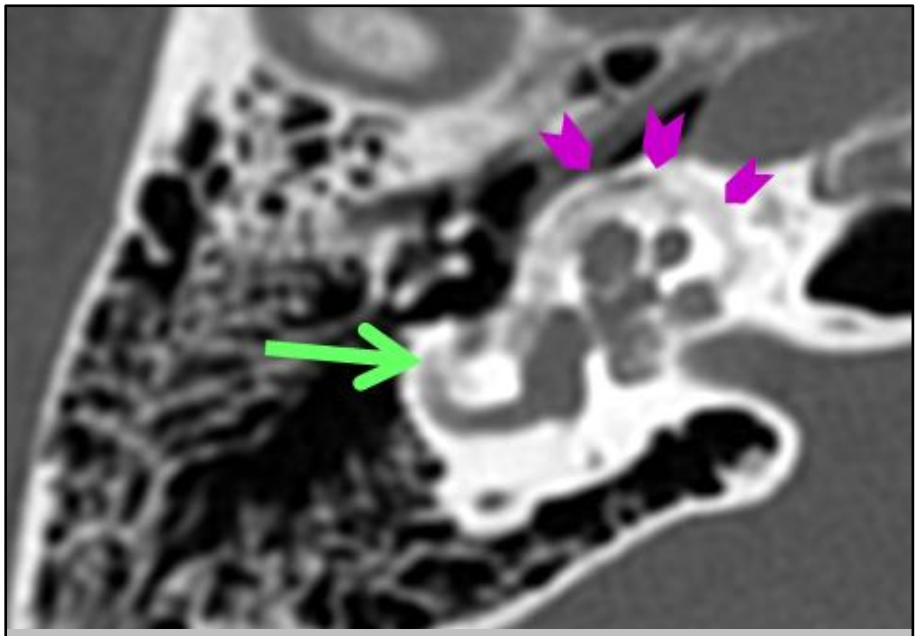


Axial CT image of the **left** temporal bone

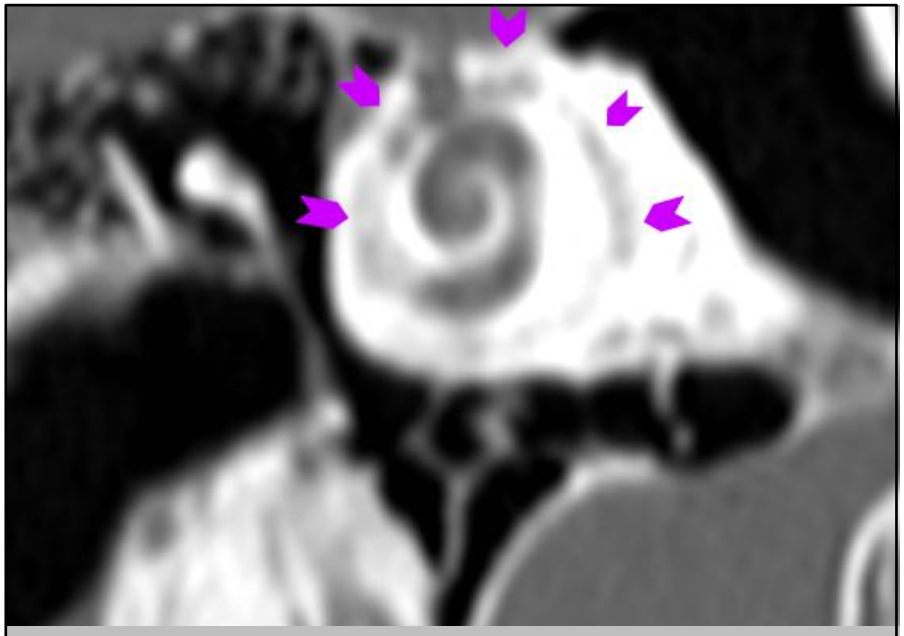
Hypodensity antero-inferior to the internal auditory canal (HAIIAC)

III. Results

Radiological evaluation CT: retrofenestral hypodensities



Axial CT image of left temporal bone
Pericochlear hypodensities
Horizontal SCC hypodensities



Coronal reformatted CT image of left temporal bone
Pericochlear hypodensities

III. Results
Association audiometry – imaging findings

Audiometric results	No. of ears	Hypodensities		
		None	Fenestral	Fenestral + retrofenestral
Normal hearing	2 ears (2 pts)	**1**/2	1/2	_
Conductive HL	4 ears (3 pts)	2/4 (EVA + S SCCD)	**2**/4	_
Mixed HL	20 ears (12 pts)	2/20 (thick slices)	3/20	**15**/20
SNHL	2 ears (2 pts)	**1**/2	_	1/2 (piston)
Deafness	5 ears (3 pts)	_	_	**5**/5

The **occurrence and location** of the hypodensities affecting various structures of middle and inner ear corresponded to the expectations based on the **type of hearing** loss in **24/33 ears** (**73%**).

III. Results

Association hearing loss severity – affected structures

- Involvement of fenestral temporal bone structures in function of magnitude ABG

Structure	ABG					
	< 15 dB	15 -25 dB	25 – 35 dB	35 – 45 dB	> 45 dB	Total
FAF	1/4	5/7	4/7	5/5	4/4	19/27*
Oval window	2/4	3/7	5/7	5/5	3/4	18/27*
Round window	1/4	3/7	4/7	4/5	3/4	15/27*

*6 ears with undeterminable ABG excluded.

- **Significant positive correlation** between the number of affected fenestral structures on CT and the magnitude of ABG on the audiogram (r=0.464; p <0.05)
- No association between magnitude of ABG and involvement of a specific structure

III. Results

Association hearing loss severity – affected structures

- Involvement of retrofenestral temporal bone structures in function of the average bone conduction threshold

Structure	Average bone conduction threshold					
	< 15 dB HL	15 -25 dB HL	25 – 35 dB HL	35 – 45 dB HL	> 45 dB HL	Total
Cochlear turns	0/6	6/12	2/4	3/5	5/6	16/33
Facial nerve canal	0/6	4/12	1/4	3/5	2/6	10/33
Semicircular canals	0/6	1/12	1/4	0/5	3/6	5/33
HAIIAC	0/6	9/12	3/4	2/5	2/6	16/33
Double ring sign	0/6	4/12	0/4	2/5	5/6	11/33

- **Significant positive correlation** between the number of affected retrofenestral structures on CT and the average bone conduction threshold determined by audiometry (r=0.471; p <0.05)
- No association between magnitude bone conduction threshold and involvement of a specific structure

IV. Discussion & Conclusion

- Similarities with otosclerosis (otospongiosis)

- **Type** of hearing loss in OI
  associated with **location** of hypodens areas:
  - Conductive loss: fenestral
  - Mixed hearing loss: fenestral + retrofenestral

- **Severity** of the conductive/sensorineural component
  associated with **extent** of hypodensities affecting temporal bone structures

- ? Severe OI (type III): higher risk on
  - EVA
  - dehiscent SCC

IV. Discussion & Conclusion

- Surplus value of temporal bone imaging in OI:
  - Confirmation of diagnosis of OI
  - Etiology of hearing loss
  - Extent of temporal bone affection
  - Unsatisfactory hearing gain after stapes surgery
  - Preoperative evaluation in stapes surgery and cochlear implantation

- Submillimetric spatial resolution recommended


THANK
YOU