

Otological and audiological findings from the Danish Study

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Background

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ORIGINAL ARTICLE



Osteogenesis imperfecta and the teeth, eyes, and ears—a study of non-skeletal phenotypes in adults

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Aim

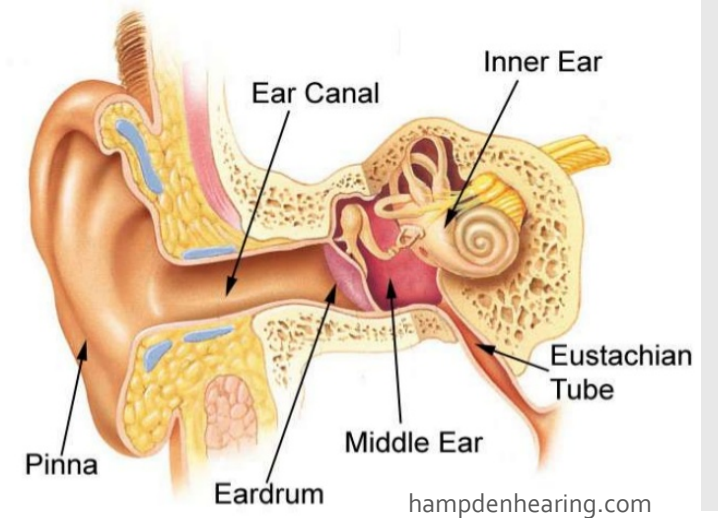
To investigate the relationship between OI type and collagen type 1 defect and hearing loss

Subjects

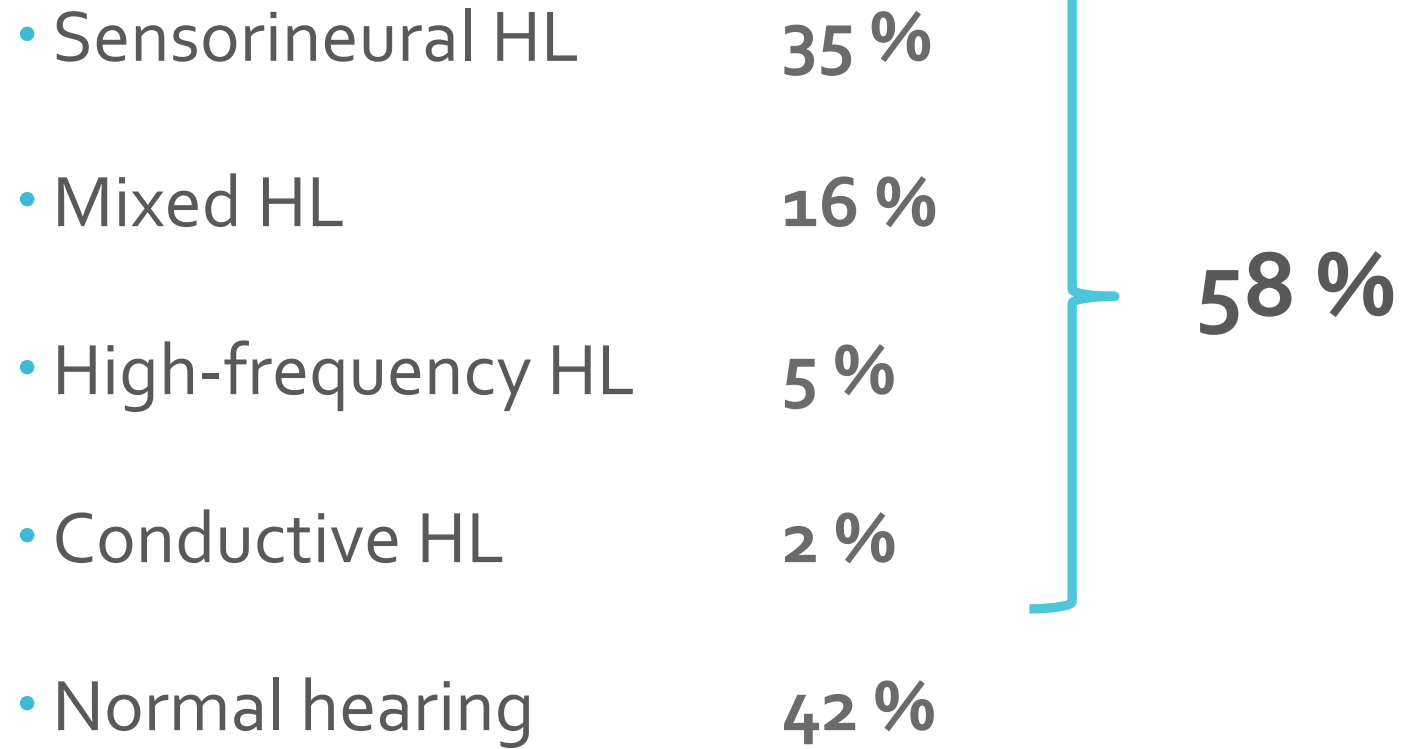
- 62 participants (124 ears)
 - 44 type I OI
 - 6 type III OI
 - 12 type IV OI
- 36 females & 26 men
- Age: 46 (20-77) years [median (range)]

Methods

- Otomicroscopic examination
- Pure-tone and speech audiometry
 - Air conduction:
0.25, 0.5, 1, 2, 3, 4, 6, and 8kHz
 - Bone conduction:
0.5, 1, 2, 3, and 4 kHz
- Hearing loss (HL) classified as:
 - Conductive
 - Sensorineural
 - High-frequency sensorineural
 - Mixed



Hearing pattern



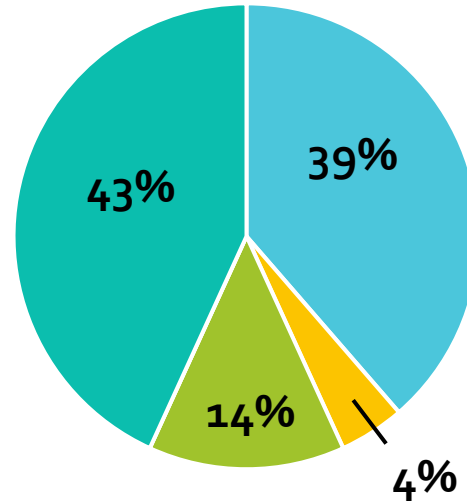
OI type and type of hearing loss

| | OI TYPE | | |
|------------------------|----------|------------|-----------|
| | I (n=88) | III (n=12) | IV (n=24) |
| SNHL | 34 (39%) | 3 (25%) | 6 (25%) |
| Mixed HL | 12 (14%) | 5 (42%) | 3 (13%) |
| High-freq. SNHL | 4 (4%) | 0 (0%) | 2 (8%) |
| Conductive HL | 0 (0%) | 3 (25%) | 0 (0%) |
| Normal hearing | 38 (43%) | 1 (8%) | 13 (54%) |

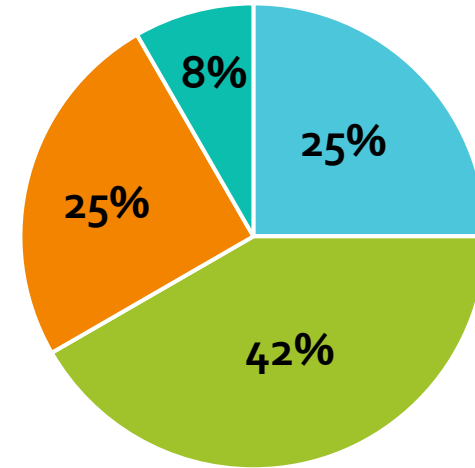
P<0.0005

OI type and type of hearing loss

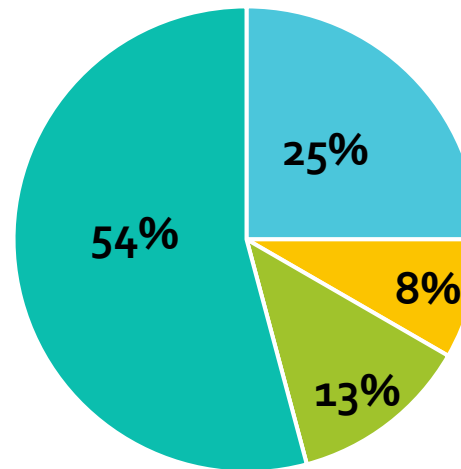
Type I
n=88



Type III
n=12



Type IV
n=24



P<0.001

■ SNHL ■ Mixed HL ■ High-freq. SNHL ■ Conductive ■ Normal hearing

OI type and hearing loss severity

| | OI TYPE | | |
|--|-----------------|-------------------|------------------|
| <i>Hearing loss severity (dB)</i> | I (n=88) | III (n=12) | IV (n=24) |
| Normal hearing (<26dB) | 53 (60%) | 1 (8%) | 18 (75%) |
| Mild HL (26-40 dB) | 20 (23%) | 6 (50%) | 4 (17%) |
| Moderate HL (41-55dB) | 5 (6%) | 2 (17%) | 0 (0%) |
| Moderately severe HL (56-70 dB) | 3 (3%) | 3 (25%) | 1 (4%) |
| Severe HL (71-90 dB) | 5 (6%) | 0 (0%) | 1 (4%) |
| Profound HL (>90 dB) | 2 (2%) | 0 (0%) | 0 (0%) |

P<0.005

OI type and collagen type 1 defect

| | OITYPE | | |
|----------------------------|----------|-----------|-----------|
| | I (n=44) | III (n=6) | IV (n=12) |
| Normal | 3 (7%) | 0 (0%) | 0 (0%) |
| Quantitative defect | 29 (66%) | 1 (17%) | 1 (8.5%) |
| Qualitative defect | 2 (4%) | 3 (50%) | 10 (83%) |
| Not available | 10 (23%) | 2 (33%) | 1 (8.5%) |

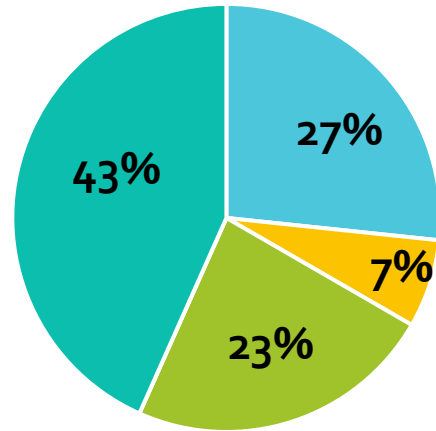
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Collagen type 1 defect and type of hearing loss

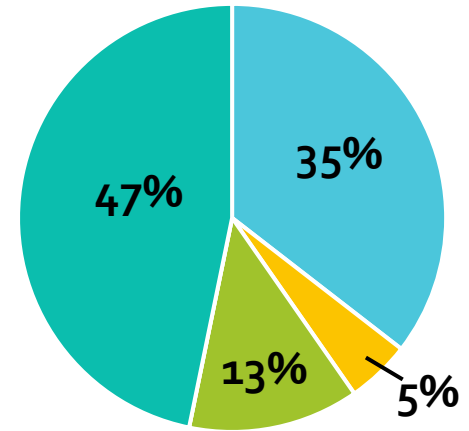
| | Collagen type 1 defect (n=98) | | |
|------------------------|-------------------------------|---------------------|--------------------|
| | Normal (n=6) | Quantitative (n=62) | Qualitative (n=30) |
| SNHL | 2 (33%) | 22 (35%) | 8 (27%) |
| High-freq. SNHL | 1 (17%) | 3 (5%) | 2 (7%) |
| Mixed HL | 0 (0%) | 8 (13%) | 7 (23%) |
| Conductive HL | 0 (0%) | 0 (0%) | 0 (0%) |
| Normal hearing | 3 (50%) | 29 (47%) | 13 (43%) |

Collagen type 1 defect and type of hearing loss

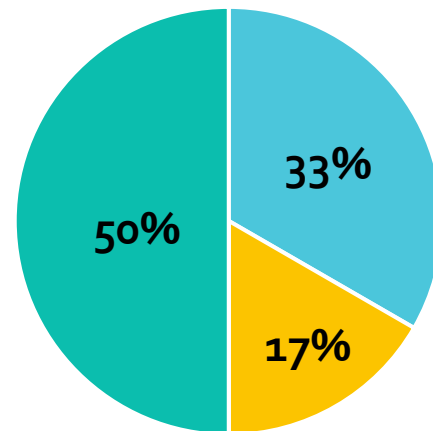
Qualitative
n=30



Quantitative
n=62



Normal collagen
n=6



P=0.6

■ SNHL ■ High-freq. HL ■ Mixed HL ■ Conductive HL ■ Normal hearing

Collagen type 1 defect and hearing loss severity

| | Collagen type 1 defect (n=98) | | |
|--|-------------------------------|----------------------------|---------------------------|
| <i>Hearing loss severity (dB)</i> | Normal (n=6) | Quantitative (n=62) | Qualitative (n=30) |
| Normal hearing (<26 dB) | 6 (100%) | 39 (63%) | 18 (60%) |
| Mild HL (26-40 dB) | 0 (0%) | 14 (23%) | 6 (20%) |
| Moderate HL (41-55 dB) | 0 (0%) | 4 (6%) | 2 (7%) |
| Moderately severe HL (56-70 dB) | 0 (0%) | 2 (3%) | 4 (13%) |
| Severe HL (71-90 dB) | 0 (0%) | 1 (2%) | 0 (0%) |
| Profound HL (>90 dB) | 0 (0%) | 2 (3%) | 0 (0%) |

OI type and otosurgery

| | OITYPE | | |
|----------------------------------|-----------------|-------------------|------------------|
| <i>Surgical procedure</i> | I (n=88) | III (n=12) | IV (n=24) |
| Type III | 4 (4.5%) | 0 (0%) | 0 (0%) |
| Stapedotomi | 11 (12.5%) | 0 (0%) | 3 (12.5%) |
| No surgery | 73 (83%) | 12 (100%) | 21 (87.5%) |

P=0.48

Treatment and HL severity

| Hearing threshold | No aid n=98 | Hearing aid n=25 | Bone-anchored hearing device n=1 |
|-------------------|----------------|---------------------|-------------------------------------|
| 0 – 20 dB | 62 | 0 | 0 |
| 21- 40 dB | 31 | 8 | 0 |
| 41 – 60 dB | 3 | 7 | 0 |
| 61 – 80 dB | 2 | 7 | 0 |
| 81 dB ≤ | 0 | 3 | 1 |

Treatment and HL severity

| Hearing threshold | No aid n=98 | Hearing aid n=25 | Bone-anchored hearing device n=1 |
|-------------------|----------------|---------------------|-------------------------------------|
| 0 – 20 dB | 62 | 0 | 0 |
| 21- 40 dB | 31 | 8 | 0 |
| 41 – 60 dB | 3 | 7 | 0 |
| 61 – 80 dB | 2 | 7 | 0 |
| 81 dB ≤ | 0 | 3 | 1 |

Conclusions

- Hearing loss is a frequent finding in patients with osteogenesis imperfecta and occurs in almost 60 % of ears
- Sensorineural hearing loss is the most frequent type of hearing loss, whereas mixed hearing loss accounts for the most severe degrees of hearing loss
- Osteogenesis imperfecta type III is related to the highest occurrence of hearing loss, whereas type I is related to the most severe degrees of hearing loss
- Degree and type of hearing loss was similar for patients with qualitative and quantitative defect collagen type 1
- Treatment of hearing loss is sub-optimal, as one-fifth of ears with significantly elevated thresholds received no treatment