

Osteogenesis imperfecta – assessment of hearing loss and otosurgical procedures

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Background

- Osteogenesis imperfecta (OI) is a genetic disorder distinguished by increased bone fragility
- Prevalence approximately 500 in Denmark
- Hearing loss of sensorineural, conductive, or mixed etiology found in 50-90% of patients
- Results of otosurgery poorer than for general population
 - 75-85% vs. 90-95% (Air-Bone Gap \leq 10 dB after stapedotomy)
- Osteogenesis imperfecta affects both ossicles, bony labyrinth, temporal bone, and cranium

Aim

To quantify hearing loss and otosurgical procedures performed in a cohort of patients with osteogenesis imperfecta

Subjects

- 62 patients (124 ears) with osteogenesis imperfecta
 - 15 examined at Odense University Hospital
 - 47 examined at Aarhus University Hospital
- 36 females & 26 males
- Age: 46 (20-77) years [median (range)]

Methods

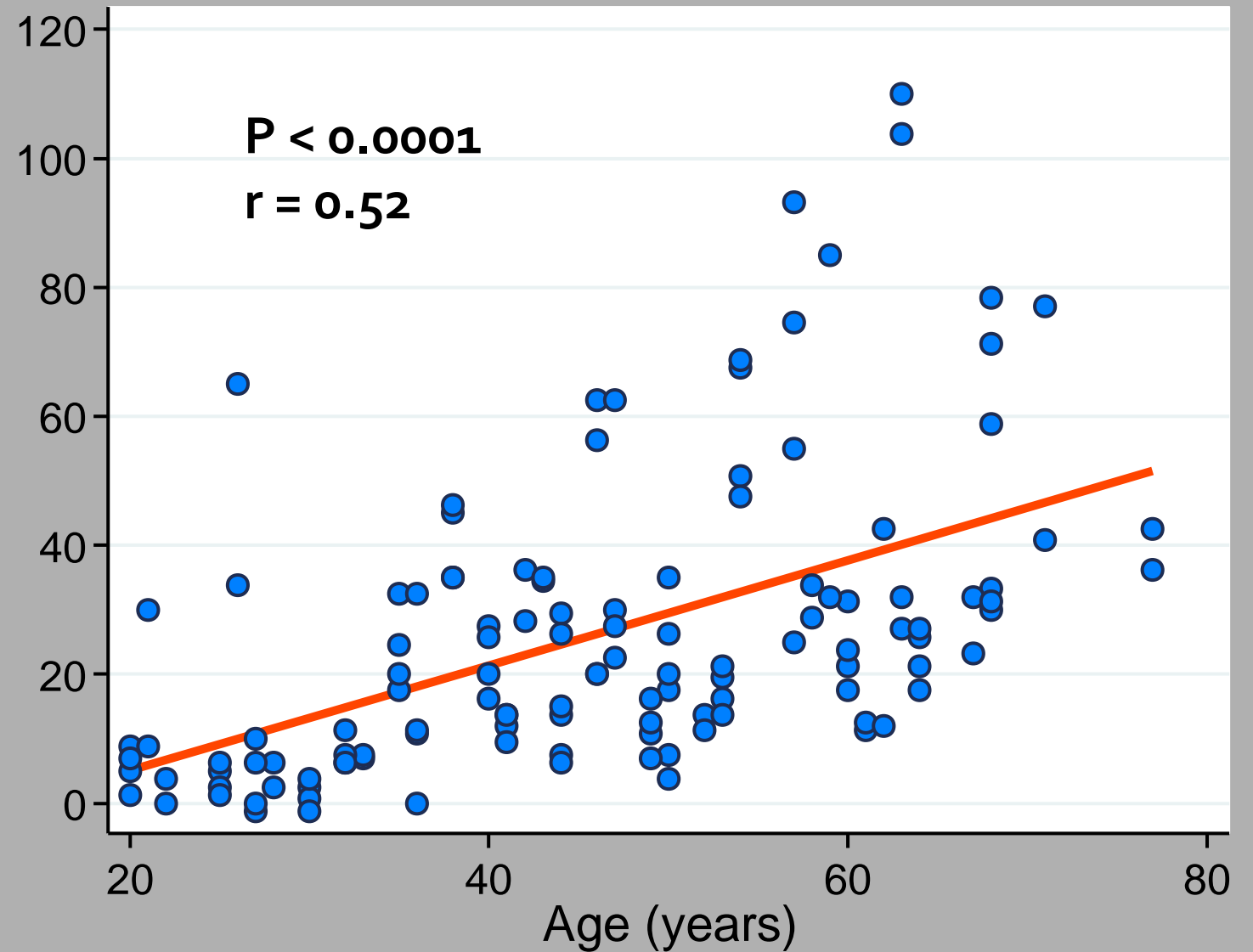
- Otomicroscopic examination
- Pure-tone and speech audiometry
 - Air conduction:
0.25, 0.5, 1, 2, 3, 4, 6, and 8 kHz
 - Bone conduction:
0.5, 1, 2, and 4 kHz
- Standardized questionnaire
- Hearing loss (HL) classified as:
 - Conductive
 - Sensorineural
 - Mixed
 - High-frequency sensorineural

Hearing pattern

• Normal hearing	42 %	
• Sensorineural HL	35 %	} 58 %
• Mixed HL	16 %	
• High-frequency HL	5 %	
• Conductive HL	2 %	

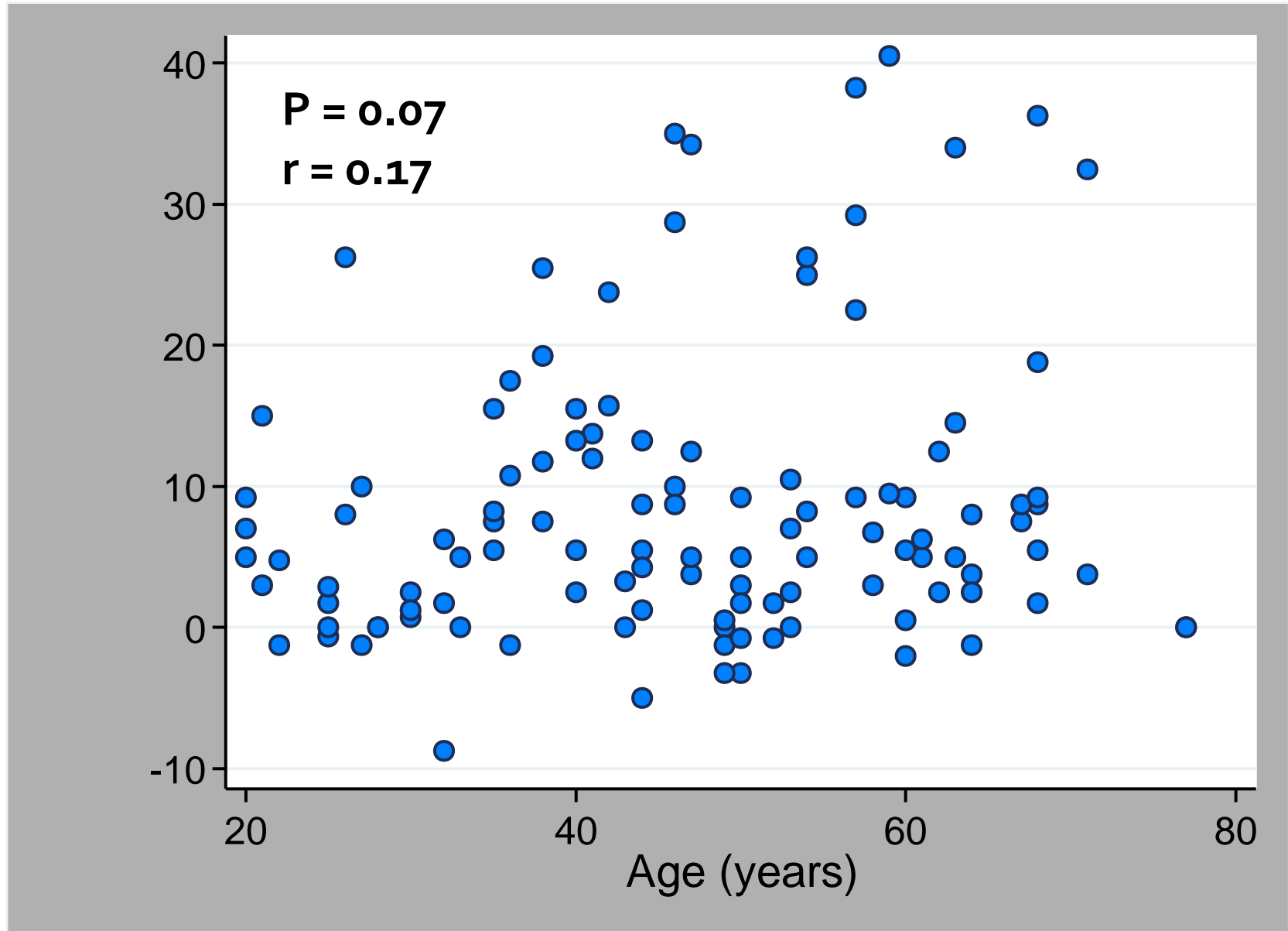
Age and hearing thresholds

Pure-Tone Average Air Conduction



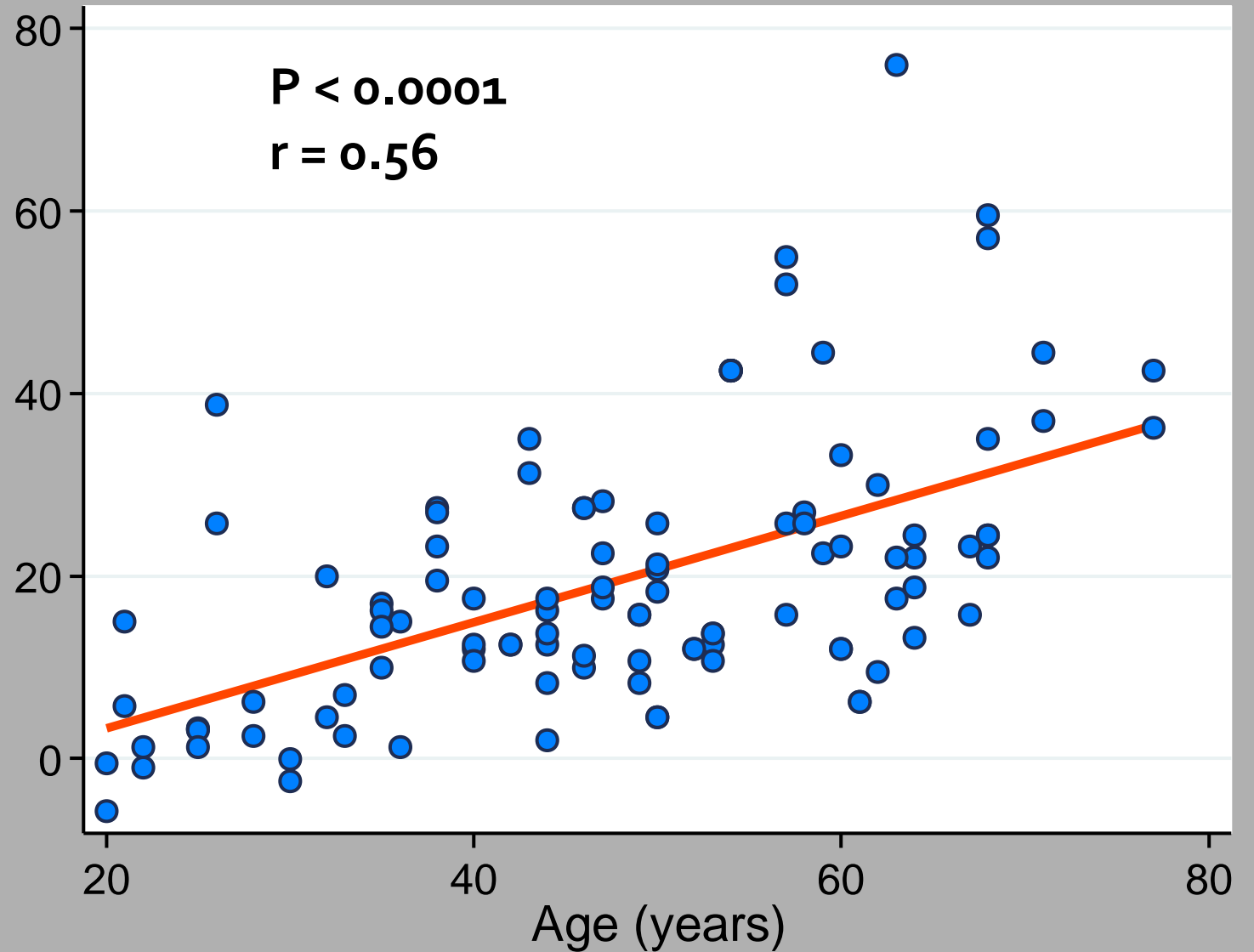
Age and hearing thresholds

Air-Bone Gap



Age and hearing thresholds

Pure-Tone Average Bone Conduction



Otosurgical procedures

Procedure	Number of ears	%
Type III tympanoplasty	4	3.2
Stapedotomy	14	11.3
No surgery	106	85.5
<i>Total</i>	<i>124</i>	<i>100</i>

Otosurgical procedures and hearing thresholds

Procedure	PTA AC (dB)	ABG (dB)
Type III tympanoplasty (n=4)	24 (20-43)	4 (2-13)
Stapedotomy (n=14)	36 (25-85)	9 (2-41)
No surgery (n=106)	18 (-1-110)	6 (-9-39)

PTA AC: Pure-Tone Average Air Conduction
ABG: Air-Bone Gap

[Median (range)]

Otosurgical procedures and hearing thresholds

Procedure	PTA AC (dB)	ABG (dB)
Stapedotomy	36 (25-85)	9 (2-41)
- Mixed (n=5)	75 (63-85)	25 (19-41)
- Sensorineural (n=9)	28 (25-59)	5 (2-15)

PTA AC: Pure-Tone Average Air Conduction
ABG: Air-Bone Gap

[Median (range)]

Conclusions

- Progression of hearing loss in osteogenesis imperfecta due to sensorineural deterioration
- Approximately one in six of adult ears affected by osteogenesis imperfecta undergo otosurgery
- Surgical outcome is acceptable