

Comparative Analysis Of Auditory Profile Of Individuals With Infratentorial Superficial Siderosis And With Age-Related Hearing Loss Using Patient-Reported Outcome Measures: A Case-Control Study

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INTRODUCTION

Infratentorial superficial siderosis (iSS) is a rare neuro-otological disorder. It results from chronic extravasation of blood into cerebrospinal fluid (often from dural defects) and deposition of iron-degradation product hemosiderin on the surfaces of CNS structures including 8th cranial nerves.^{1,2} iSS is likely to affect individuals in their 2nd half of life.

its most common feature is progressive hearing loss (HL).

Pure-tone audiometry (PTA) often shows high-frequency sensorineural HL resembling age-related hearing loss (ARHL) pattern³

Hearing-specific patient reported outcome measures (PROMs) have been used to identify the auditory profile in various patient groups (such as with auditory-processing disorder, stroke or ARHL).⁴

AIM

To compare the auditory profile of individuals with iSS and with ARHL using hearing-specific PROMs

METHODS

- Study received permission from UCL Research Ethics Committee (UCL REC 17413/001)
- We conducted anonymous online case-control study using Research Electronic Data Capture platform REDCap for survey delivery
- **Recruitment:** we contacted relevant charities, organisations and patient groups inviting individuals ≥18 years old with diagnosis of iSS or ARHL to participate in the survey, between 20/4/2020 and 31/7/2021
- Following consent, eligible individuals were provided with study-specific questions and PROMs (**Table 1**)
- Statistical analysis was performed using SPSS (v26-28, IBM Armonk NY)

	mAIADH ⁵	SSQ ⁶	ERSA ⁷	TFI ⁸
Domains	5	3	3 (+1)*	8
Intelligibility of speech:	Speech	Speech	Effect of hearing on:	Intrusiveness
• in quiet	Spatial	• Quality of life	Sense of control	
• in noise	Other Qualities of hearing	• Personal life	Cognitive effect	
Sound:		• Social life	Sleep	
• detection		• Occupational life	Auditory function	
• recognition			Relaxation	
• localisation			Quality of life	
			Emotional distress	
Items	28	49	15+5*	25
Outcome measures	Total score	Total score	Total score /150*	Total score
Value range (worst-best)	0 to 3 (average score)	0 to 10	0 to 50 (domain)	100 to 0
	0 to 84 (sum)		0 to 150 (total)*	

Table 1. Characteristics of PROMs used. Legend: mAIADH modified Amsterdam Inventory for Auditory Disability and Handicap; SSQ Speech, Spatial and other Qualities of hearing; ERSA Evaluation du Retentissement de la Surdit  chez l'Adulte (Evaluation of the Impact of Hearing Loss in Adults); TFI Tinnitus Functional Index. *'Occupational life' domain not included in the total score due to low completion rate.

RESULTS

Participant demographics	iSS group (n=47)	ARHL group (n=30)
Gender (males, %)	28 (60)	12 (40)
Age at survey (median, IQR), years	59 (15)	75 (10)
Age at onset of hearing problems (median, IQR), years	47 (21)	61 (11)
Duration of hearing problems (median, IQR), years	9 (12.5)	10 (11.3)

Table 2. Participant demographics.

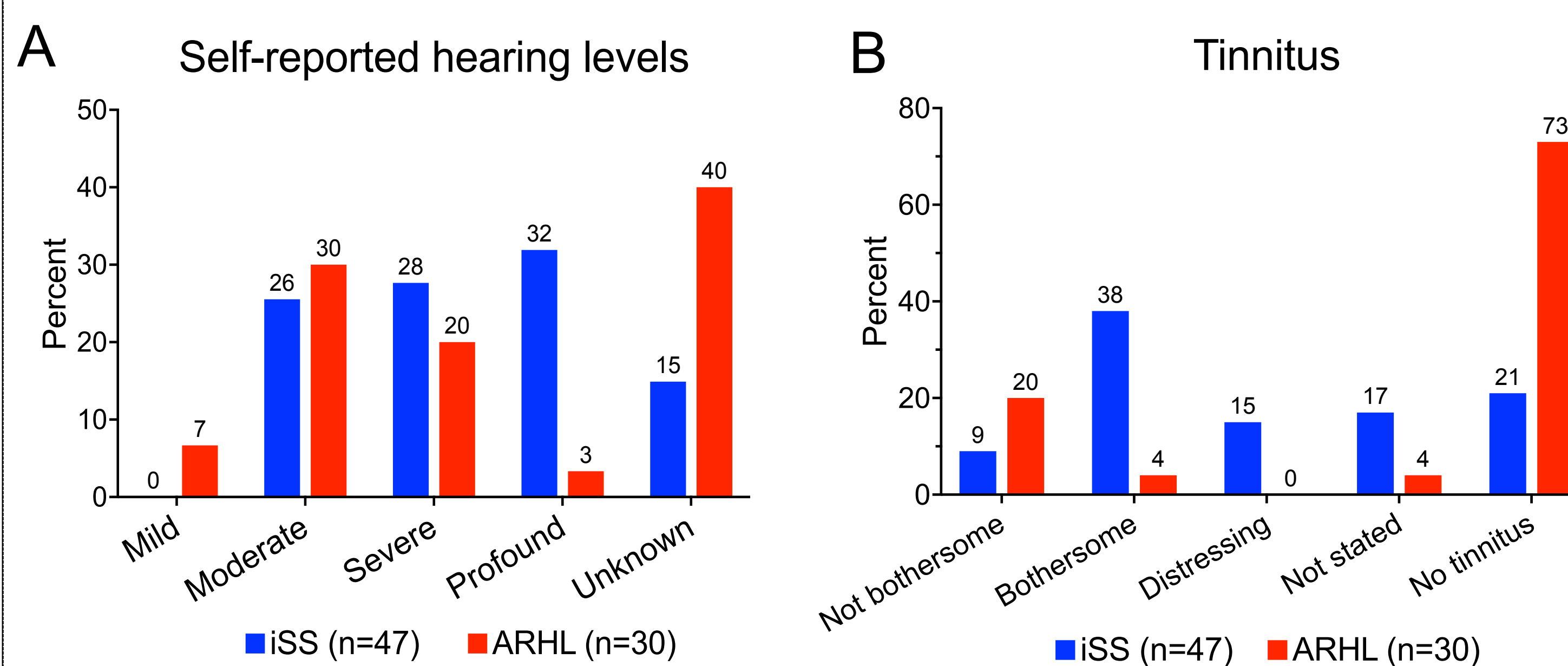


Figure 1. Participant-reported hearing levels (A) and tinnitus (B).

Participant characteristics	Mean ranks iSS group	Mean ranks ARHL group	Z-value	p-value
Age	32.3	49.4	3.27	0.001**
Duration of hearing symptoms	36.5	40.3	0.74	0.458
Hearing levels	33.6	20.5	-2.88	0.004**
Tinnitus severity	20.7	9.4	-2.69	0.007**

Table 3. Between-group comparison of participant characteristics using non-parametric Mann-Whitney U test; alpha level set at 0.05. **p-value significant at <0.01.

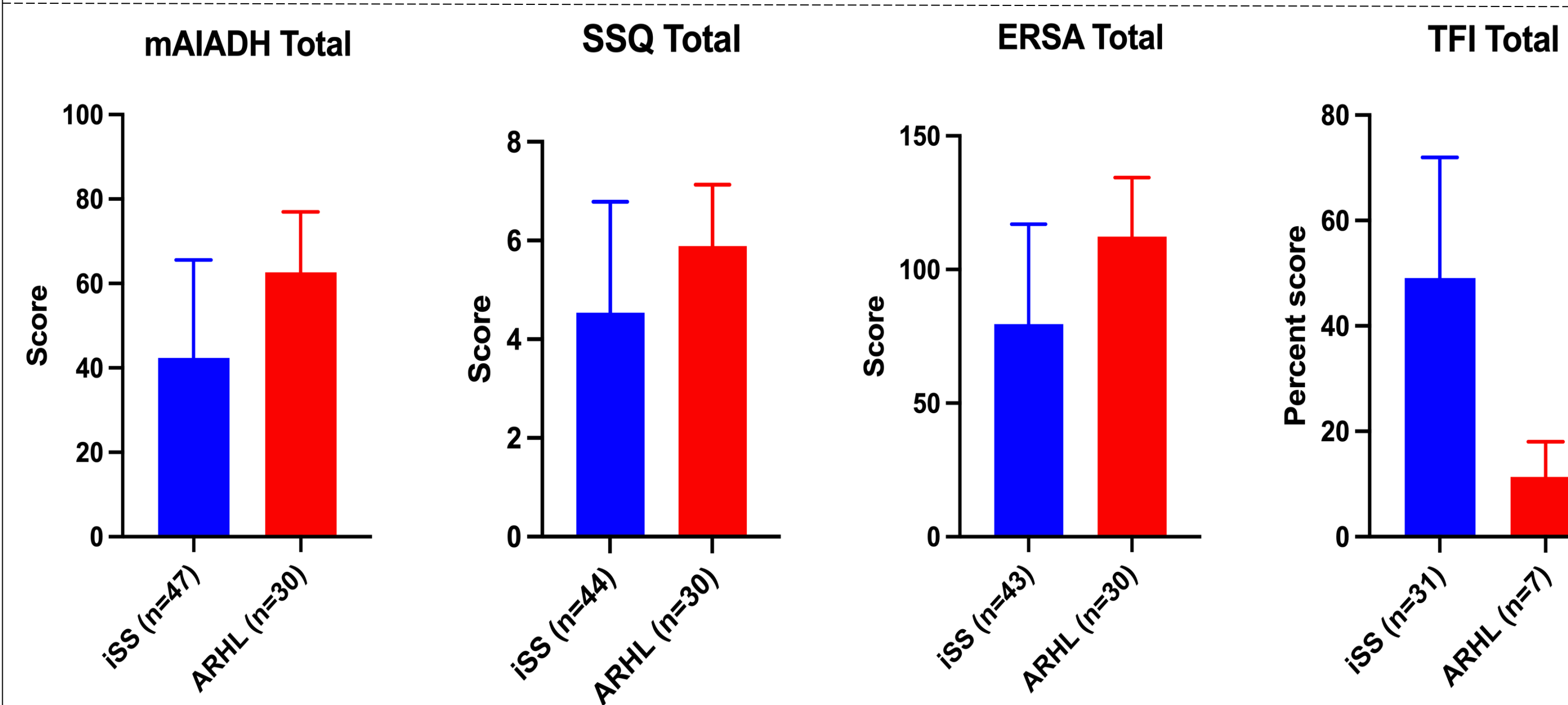


Figure 2. Box plot charts with error bars for iSS and ARHL groups with total scores for each PROM. Mean and standard deviation provided.

mAIADH	Quade's ANCOVA		SSQ	Quade's ANCOVA	
	F-score	p-value		F-score	p-value
Speech in noise	1.07	0.307	Speech	0.523	0.473
Speech in quiet	1.88	0.176	Spatial	0.999	0.322
Sound localisation	4.182	0.046*	(Other) Qualities	0.439	0.511
Sound recognition	19.09	<0.001**	TOTAL	1.115	0.296
Sound detection	2.90	0.094	TFI	Quade's ANCOVA	
TOTAL	3.18	0.080		F-score	p-value

ERSA	Quade's ANCOVA		TOTAL (/150)	Quade's ANCOVA	
	F-score	p-value		F-score	p-value
Quality of life	8.24	0.006**	TOTAL	12.54	0.001**
Personal life	0.97	0.330	Intrusiveness	11.67	0.002**
Social life	4.78	0.033*	Sense of control	7.30	0.011*
Occupational life	8.17	0.014*	Cognitive	4.0	0.053
TOTAL (/150)	5.10	0.028*	Sleep	13.99	0.001**
			Auditory	10.70	0.002**
			Relaxation	6.96	0.012*
			Quality of life	6.343	0.017*
			Emotional distress	1.92	0.175

Table 4. Between-group comparison of participant scores, using non-parametric Quade's ANCOVA controlling for hearing levels. Alpha level set at 0.05; *p-value significant at 0.05; **p-value significant at 0.01.

CONCLUSION

Our study demonstrated that:

- individuals with iSS-related HL are likely to be younger than with ARHL
- iSS-related HL is likely to be worse and of earlier onset than ARHL
- Hearing impairment in both groups is affected by presence of noise, reflected in low scores in mAIADH Speech-In-Noise domain
- iSS-related hearing impairment in mAIADH domains of Sound Localisation and Sound Recognition appears worse in individuals with iSS and may be a distinguishing feature of iSS-related HL as compared to ARHL
- Tinnitus appears more prevalent and of greater severity in individuals with iSS, likely to be reflected in worse TFI scores (than for ARHL group)
- Negative impact of hearing impairment on personal life and social life was indicated by low scores in both groups for these domains (ERSA)

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