Hearing aid fitting for visual and hearing impaired patients with Usher syndrome type IIa

Hartel, B.P.,*† Agterberg, M.J.H.,*‡ Snik, A.F.,*‡ Kunst, H.P.M.,*§ van Opstal, A.J.,‡ Bosman, A.J.*†[a] & Pennings, R.J.E.*§[a]

*Department of Otorhinolaryngology, Radboud University Medical Center, †Radboud Institute for Health Sciences, Radboud University Medical Center, ‡Department of Biophysics, Donders Institute for Brain, Cognition and Behaviour, Radboud University Nijmegen, §Donders Institute for Brain, Cognition and Behaviour, Radboud University Nijmegen, Nijmegen, The Netherlands

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Objectives: Usher syndrome is the leading cause of hereditary deaf-blindness. Most patients with Usher syndrome type IIa start using hearing aids from a young age. A serious complaint refers to interference between sound localisation abilities and adaptive sound processing (compression), as present in today's hearing aids. The aim of this study was to investigate the effect of advanced signal processing on binaural hearing, including sound localisation.
Design and participants: In this prospective study, patients were fitted with hearing aids with a nonlinear (compression) and linear amplification programs. Data logging was used to objectively evaluate the use of either program. Performance was evaluated with a speech-in-noise test, a sound localisation test and two questionnaires focussing on self-reported benefit.
Results: Data logging confirmed that the reported use of hearing aids was high. The linear program was used significantly more often (average use: 77%) than the nonlinear program (average use: 17%). The results for speech intelligibility in noise and sound localisation did not show a significant difference between type of amplification. However, the self-reported outcomes showed higher scores on 'ease of communication' and overall benefit, and significant lower scores on disability for the new hearing aids when compared to their previous hearing aids with compression amplification.
Conclusions: Patients with Usher syndrome type IIa prefer a linear amplification over nonlinear amplification when fitted with novel hearing aids. Apart from a significantly higher logged use, no difference in speech in noise and sound localisation was observed between linear and nonlinear amplification with the currently used tests. Further research is needed to evaluate the reasons behind the preference for the linear settings.

Usher syndrome (USH) is the leading cause of hereditary deaf-blindness. This autosomal recessively inherited disorder is characterised by sensorineural hearing impairment, retinitis pigmentosa (RP) and in part of the cases vestibular dysfunction. USH is clinically and genetically heterogeneous and has a prevalence of 4.4–6.2 per 100.000 inhabitants.[1–3] Usher syndrome type II is one of the three clinical types of USH, and Usher syndrome type IIa (OMIM276901) is the most common genetic subtype, accounting for more than half of the USH patients.[4–6] Pathogenic mutations of this subtype are identified in the *USH2A* gene located on chromosome 1q41.[7,8] Patients with Usher syndrome type IIa have a congenital moderate to severe high-frequency hearing impairment, intact vestibular function and RP, a progressive retinal degenerative disease that usually first becomes manifest in the second decade of life and eventually leads to blindness.

Most patients with Usher syndrome type IIa use hearing aids from a young age.[9] During their lives, these patients will therefore face multiple hearing aid fitting procedures. Owing to the multiple complex settings and programs of today's sophisticated hearing aids, fitting periods will be prolonged to find the best settings for these double sensory-impaired patients as described by patients with this specific syndrome.[10] Their report pointed out the difficulties experienced during hearing aid fitting and hearing aid use, which is the motivation for this study. The additional onset of visual impairment in young adulthood is thought to play a major

Correspondence: B.P. Hartel, Department of Otolaryngology, Radboud University Medical Center, Philips van Leydenlaan 15, P.O. Box 9101, Nijmegen 6500 HB, The Netherlands. Tel.: +31 (0)24 3617203; fax: +31 (0) 24 3540251; e-mail: bas.hartel@radboudumc.nl
[a]Shared last authors.

role in the lengthy fitting procedures. The impact of the hearing disability is probably more severe because of the additional effects of RP, like the loss of visual feedback.

Nowadays, hearing aids are equipped with advanced algorithms focusing on the largest consumer group: the elderly. Many algorithms have been developed to optimise speech intelligibility and to provide comfortable hearing. However, less attention is given to the preservation of natural cues used for localisation, such as interaural time difference (ITD) and interaural level difference (ILD); especially in the horizontal (azimuth) plane, the difference in sound arrival at both ears (ITD) for the lower frequencies (<1.5 kHz) and the difference in sound level between both ears (ILD) for the higher frequencies (>3 kHz) are important for sound localisation.[11]

The aim of this study was to investigate whether advanced signal processing of sound by hearing aids has a positive or negative effect on speech intelligibility in noise (with spatially separated speech and noise sources) and on sound localisation. Both tasks require binaural hearing abilities. The patients were fitted with novel hearing aids with a nonlinear (compressive) and linear amplification program. It was hypothesised that the linear program affects sound localisation minimally because the ITD and ILD-cues are potentially less perturbed.[12] This may lead to better sound localisation performance when compared to the nonlinear program. In contrast, the nonlinear program may provide better audibility and speech recognition in noisy environments, when compared to a linear fitting.[13] To test these hypotheses, details of hearing aid use were retrieved from data logging and speech reception in noise, and sound localisation was tested in an elaborate set-up.[14] Furthermore, two questionnaires and a diary were used to evaluate the subjective benefit and reported use in daily life of the newly fitted hearing aids.

Patients and methods

Patients

The patients with Usher syndrome type IIa were extracted from the Nijmegen Usher syndrome database. Patients were included if they were clinically diagnosed with Usher type II, had two identified pathogenic mutations in *USH2A*, had a pure-tone average (0.5–8 kHz) better than 80 dB HL and were above 18 years of age. Twenty-four adults were selected for participation and contacted. Finally, eighteen of them decided to participate in this study. Six patients refused without specified reasons. Some of the patients participated in former studies on hearing in Usher syndrome type IIa from our centre.[15,16]

This study was approved by the local ethics committee (nr. 2012/520).

Audiometric evaluation and hearing aids

At the first visit, pure-tone air and bone-conduction thresholds were assessed for frequencies ranging from 0.25 to 8 kHz according to the ISO 8253-1 standard [ISO 8253-1]. All patients were bilaterally fitted with Phonak Naida Q SP hearing aids (Phonak AG, Stäfa, Switzerland), hereafter referred to as 'new hearing aids'. The fitting was performed using the Target system 3.1. In six patients', ear-moulds were replaces with the standard occluding ear-moulds. These patients could adapt at least 2 weeks with the Phonak hearing aids and a nonlinear program to get used to the ear-moulds before the tests started. The vent diameter varied according to hearing loss, but all 18 patients were fitted with vents smaller than 2 mm.

Two listening programs were activated, and for 13 patients, a telecoil program was added. The first program consisted of a nonlinear fitting on the basis of the NAL-NL2 rule.[18] Minor adjustments in gain were made according to remarks from the patients. The nonlinear program used syllabic compression (SC) with an attack time of 10 ms and a release time of 80 ms. In this program, microphone directionality was set as a static, input independent, beam former ('Real-Ear sound'; to compensate to some extent for pinna function) and the default adaptive features were activated. The linear program was adapted to the nonlinear program by setting the same gain for a 65 dB SPL input. A compression ratio of one was aimed for as allowed by the patient's dynamic range, and all adaptive features were deactivated, apart from feedback suppression, which was set to medium. Microphone directionality was fixed to omnidirectional.

Figure 1 shows that the CR in the linear program [mean 1.2, standard deviation (SD) 0.2] was significantly lower at all frequencies compared to the CR in the nonlinear program (mean 1.8, SD 0.4) (panel a). The mean CR for the linear program was not 1.0 because of the reduced dynamic range in the higher frequencies. This is illustrated in panel b, in which a slight increase in CR can be seen in the higher frequencies at input levels between 65 and 80 dB SPL.

When the hearing aids were switched on, half of the patients started with the linear program, whereas the other half started with the nonlinear program. Wireless communication between both hearing aids was activated (Quick-Sync) to enable simultaneous change of sound level and program across ears. Volume control was available but at follow-up, none of the participants reported to have daily changed the volume. All patients were instructed to try both programs in all situations. Between the two test sessions, all patients were contacted and five patients were subsequently

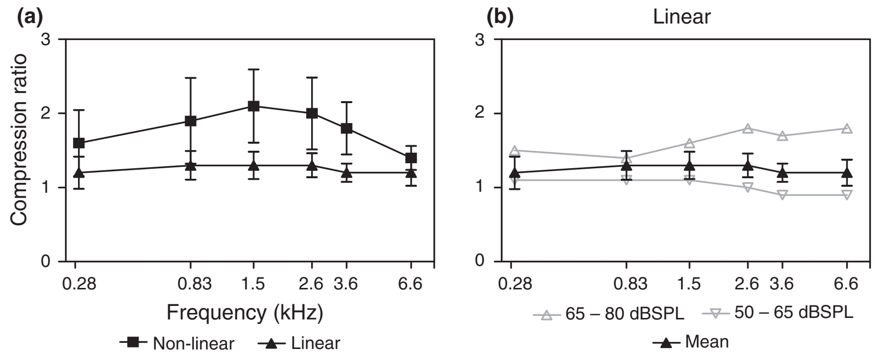

Fig. 1. Compression ratios for the linear and nonlinear program. (a) The mean compression ratios in the nonlinear program are represented by squares and continuous line with the standard deviation on either side of the squares. The mean compression ratios in the linear program are represented by the triangles and continuous line with the standard deviation on either side of the triangles. (b) Compression ratios for the linear program between 50–65 dB SPL and 65–80 dB SPL. The mean compression ratios in the linear program are represented by black triangles and continuous black line with the standard deviation on either side of the triangles. The average compression ratios in the linear program between 50 and 65 dB SPL are represented by the grey downward triangles and continuous grey line. The average compression ratios in the linear program between 65 and 80 dB SPL are represented by grey upward triangles and continuous grey line.

referred to their own hearing aid dispenser for minor (documented) adjustments of gain, only for the nonlinear program. At follow-up, the overall use, individual program use and changes in volume were retrieved from the new hearing aids. Moreover, data logging was activated to retrieve the number of days, hours and used programs.

Speech intelligibility

Speech intelligibility was measured in silence and in noise with the patient's own hearing aids before fitting and with their new hearing aids with either program (nonlinear and linear) directly after fitting and at follow-up. The speech test used was the Dutch matrix test[19] in an open-set response format, in which listeners had to repeat the words they understood from target sentences spoken by a female. These sentences were always presented from the front (at 1 m), and noise was either presented from the front or randomly from +90 or −90 degrees (at 1 m, S_0/N_{s90} and S_0/N_{f90}). The stationary noise had an average power spectrum equal to that of the sentences.[19] Additionally, a single-talker male babble noise was used based on the International Speech Test Signal.[20,21] The level of noise was adaptively varied according to the Brand and Kollmeier procedure,[22] with a minimum step size of ±1 dB. In the measurement of speech intelligibility in silence, the same set-up was used without noise and the first word was presented at 65 dBA. Two lists of ten sentences were presented per noise configuration and listening condition and the outcomes averaged to limit the intra-individual variation. Two training lists were used to familiarise subjects with the task, the first starting with an easy speech level of 65 dBA without noise. The second training list was presented with a 15 dB SNR (target speech at 65 dBA, noise at 50 dBA). Noise started 2 s before and ended 2 s after the target sentence presentation. The speech level was always held constant at 65 dBA. The 50% speech reception threshold (SRT) was determined over the last 7 reversals.

Sound localisation

Apparatus. The patients were seated in a comfortable chair in the centre of a completely dark and sound-attenuated room. The ambient background noise level in the room was 30 dBA. Horizontal head movements were recorded using the magnetic search coil induction technique. Patients wore a lightweight spectacle frame on which a small coil was mounted.[14] They were asked to turn their head as quickly and as accurately as possible in the perceived stimulus direction. The patient controlled stimulus onset by pressing a hand-held button while facing straight ahead.

Stimuli. The stimuli were digitally generated in Matlab (R2012a, The Mathworks, Inc., Natick, MA, USA). The sound was presented by a broad-range loudspeaker (MSP-30; Monacor International GmbH, Bremen, Germany). Fifty-eight loudspeakers were mounted on a vertical hoop at 100 cm of the patient that could turn to every position in azimuth. Three different acoustic stimuli were presented; a broadband stimulus (0.5–20 kHz; BB), a low-pass filtered noise stimulus (LP; high-frequency cut-off at 0.5 kHz) and a high-pass filtered noise stimulus (HP; low-frequency cut-off at 3 kHz). For the BB and HP stimuli, sound intensities were roved from 55 to 75 dBA in steps of 10 dB. This was carried out to prevent the use of perceived loudness as a cue for localisation.[23] For a more detailed description of set-up and stimuli, see reference[14].

Experiments. A visual calibration experiment was first run to map the head-position data to known spatial locations and to demonstrate that patients had no motoric problems to direct their head to the stimulus positions. After this calibration run, 10 practice trials were presented to become accustomed to the sounds and the open-loop head-movement response procedure.

For the localisation experiment, the patient was asked to orient towards 12 LP stimuli and 36 BB and 36 HP stimuli in each condition (own hearing aids, new hearing aids nonlinear, new hearing aids linear). The stimuli were presented at randomised locations between −75 and 75 degrees in azimuth with a minimum of 20 degrees between consecutive stimuli and at zero degrees elevation. Regular breaks were introduced to prevent fatigue and to motivate the patients.

Self-reported outcomes

In addition to the objective outcome measures, two questionnaires and a diary were used to assess each patient's reported benefit, satisfaction and use of the new hearing aids.

The Abbreviated Profile of Hearing Aid Benefit (APHAB) is a hearing related benefit questionnaire.[24] It contains 24 items and is a disability-based inventory to document the outcome of hearing aid fitting and to evaluate fitting over time. The questionnaire yields scores on four subscales: ease of communication, listening under reverberant conditions, listening in background noise and aversiveness of sounds. The APHAB was used to qualify the disability and the differences in disability between hearing aids. The maximum disability was represented by a 100% of the time that certain situation occurred, the patient felt disabled, and the minimum (best score) was 0% (never disabled).

The Glasgow Hearing Aid Difference Profile (GHADP) was designed to evaluate patient reported hearing disability, handicap, hearing aid use, benefit, residual disability and satisfaction.[25] This inventory provided eight possible environments. Four of them were predetermined, and four others could be added by the patient. This questionnaire was used to address the differences between the patient's own and new hearing aids.

A non-validated diary was provided to the patient for the first week after fitting and the last week before the follow-up visit. Questions about the number of changes between programs, use of programs and satisfaction on a 1–10 Likert scale were included. Finally, patients were also asked to describe situations in which they used a specific program.

Data analysis

Patient characteristics were compared using Student's *t*-test or their nonparametric counterpart if the data deviated from a normal distribution. All analyses were performed using Prism 5.03 software (GraphPad, San Diego, CA, USA). A paired *t*-test was performed for comparison of compression ratios between programs. Binomial distribution statistics were used for simultaneous assessment of the CR over all six audio frequencies. Simultaneous significance was accepted if such differences were significant at two of more frequencies ($P < 0.05$ tail probability in the binomial distribution with $N = 6$, $P = 0.05$, $q = 0.95$). Furthermore, one-way analysis of variance (ANOVA) with *post hoc* Bonferroni correction was used to compare speech intelligibility and self-reported outcomes between the two fittings and conditions. A general significance level of $P = 0.05$ was applied in all separate tests.

Sound localisation. All responses were analysed with Matlab for each patient and condition (R2012a, The Mathworks, Inc.). The best linear fit (least squares criterion) of the stimulus–response relationship on the azimuth data was determined with the following equation:

$$\alpha_r = b + G\alpha_S$$

in which α_r is the response azimuth in degrees, b the response *bias* in degrees, G the response *gain* (dimensionless) and α_S the presented stimulus azimuth in degrees.[26] From the regression, we calculated the coefficient of determination (r^2) of the fit, as well as the mean absolute error (MAE; in degrees). A good performer should produce a *gain* and r^2 close to 1.0 and a *bias* and MAE close to 0.0 degrees. Results for each condition were averaged across patients, and the *gain changes* were normalised to enable a direct and unbiased comparison between conditions as described by Zwiers *et al.*[27]. The normalisation of the gain change was obtained with the following equation:

$$G_E = |G/G_C - 1|$$

in which G is the measured *gain* for a particular condition in the patient and G_C the *gain* for the control condition in the patient. A value of $G_E = 0$ indicates that the measured gain was equal to the control value, that is no change in gain. The absolute value ensures that systematic overshoots and undershoots yielded similar measures.[27]

Results

Patients

A total of 18 patients (nine male and nine female) with a mean age of 38.8 years (range 20–55) were included, and Table 1 shows their general characteristics. Most patients reported a childhood onset of hearing impairment and an almost lifetime use of hearing aids. All patients but one used their own hearing aids during at least 1 year.

The median follow-up period was 6.3 weeks (range 5.3–16.9).

Audiometric evaluation

A symmetrical, high-frequency, sensorineural hearing loss was observed in all patients. The mean audiogram, obtained from thresholds of the best hearing ear, is shown in Fig. 2. The Loudness Discomfort Levels (LDL), relative to the mean thresholds, clearly demonstrate a reduction in dynamic range of hearing, most pronounced at the high frequencies. This influences the calculated compression ratios, as shown in Fig. 1.

Hearing aids

All patients used their new hearing aids on a daily basis as shown in Table 2 with a mean overall use of 11.6 h/day. All but one patient kept using the new hearing aids after the study ended. That one patient preferred his old hearing aids. All patients reported to have tried the two programs in different situations. Overall, patients used the linear program on average 77% of the time (range 56%–99%), whereas this was only 17% (range 1%–44%) for the nonlinear program (two-sided; $P > 0.001$; 95% Confidence Interval (CI) 46.0–75.2). This difference was statistically significant (bold values in Table 2). No difference was noted between subgroups based on program at starting up.

Table 1. Patient characteristics

Number of patients, *n* (m/f)	18 (9/9)
Mean age, years (range)	38.8 (20–55)
Median age start hearing loss, years (range)	0 (0–5)
Median age first hearing aid, years (range)	4 (2–36)
Mean use own hearing aids, years (SD)	3.8 (1.7)
Median follow-up, weeks (range)	6.3 (5.3–16.9)

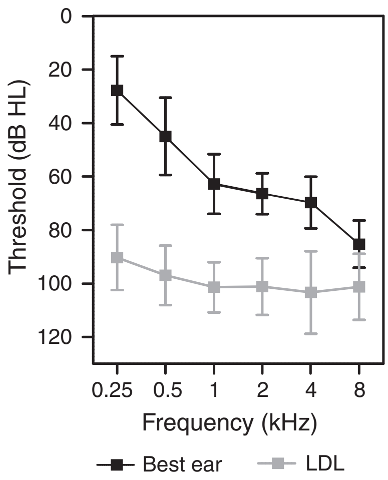

Fig. 2. Average thresholds and Loudness Discomfort Levels of the best ear. Average thresholds are represented by black squares and continuous line. Per frequency, the standard deviations are represented by thick black lines on either sides of the squares. The loudness discomfort levels are represented by the grey squares and continuous line. Per frequency, the standard deviations are represented by thick grey lines on either side of the squares. Abbreviations: LDL, Loudness Discomfort Levels; HL, hearing level.

Table 2. Data logging from the new hearing aids

Mean data logging, days (SD)	53.7 (18.0)
Mean use new hearing aids/day, hours (SD)	11.6 (4.7)
Mean use linear program, % (range)	**77 (56–99)**
Mean use nonlinear program, % (range)	**17 (1–44)**
Mean use telecoil program, % (range)	6 (0–28)

In bold, the values which differed significantly between the linear and nonlinear program.

Speech in noise

Table 3a shows no significant differences in mean SRT between the patient's own and new hearing aids with either the linear or nonlinear program. At the second session, a decrease in SRT was seen in patients with new hearing aids in both programs compared to the first measurement directly after the fitting, but this difference was not significant.

Likewise, no significant differences were found in mean SRT in noise between both programs at either visit. However, a significant improvement was seen with the own and new hearing aids (in either program) when the noise (stationary or babbled noise) was presented at −90 or +90 degrees (Table 3b).

Sound localisation

For four patients, we could not obtain localisation performance with their own hearing aids: one patient did not use

Table 3. (a) Mean SRT values for the own and new hearing aids without noise. (b) Mean signal-to-noise ratio values for the own and new hearing aids

		New nonlinear		New linear	
(a)	Own	At fitting	Follow-up	At fitting	Follow-up
SRT, mean (SD)	39.4 (4.6)	38.9 (4.6)	37.4 (4.7)	42.3 (4.2)	40.6 (4.9)

		New nonlinear		New linear	
(b)	Own	At fitting	Follow-up	At fitting	Follow-up
SNR S_0/N_{s0}, mean (SD)	−4.1 (1.6)	−3.2 (1.5)	−4.2 (1.6)	−3.8 (1.2)	−4.8 (1.8)
SNR S_0/N_{s90}, mean (SD)	−7.4 (4.3)*	−8.1 (3.3)*	−9.9 (3.9)*	−7.9 (3.5)*	−9.6 (3.7)*
SNR S_0/N_{f0}, mean (SD)	n.p.	n.p.	−6.9 (3.5)	n.p.	−6.3 (3.6)
SNR S_0/N_{f90}, mean (SD)	n.p.	n.p.	−11.0 (4.6)*	n.p.	−12.2 (4.4)*

(a) Abbreviations: SRT, speech reception threshold.
(b) The signal-to-noise ratio is the ratio at which the SRT is 50%.
Abbreviations: SNR, signal-to-noise ratio; S, Signal; N_s, Speech noise; N_f, male babble noise; N_0, 0 degrees; N_{90}, 90 degrees.
*SNR S_0/N_s (or $_f$) $_{90}$ significantly better compared to SNR S_0/N_s (or $_f$) $_0$.

his hearing aids, for one patient the hearing aids failed and for two patients the set-up failed. Figure 3 shows two typical results of sound localisation for broadband stimuli for two of the patients (#6 and #16). Patient #16 localised well with his/her own hearing aids, as well as with the two programs for the new hearing aids. Note that the bias decreased from −13 deg (with own hearing aids) to nearly 0 deg (new hearing aids). Patient #6 localised much better with the new hearing aids for either program, when compared to the own hearing aids.

Figure 4 shows all the measured values for G, r^2, b and MAE for the three conditions with broadband sounds. The results for patients #6 and #16 are highlighted. On an individual level, the good performers with their own hearing aids seem to perform equally well in all three conditions. However, the poor performers with their own hearing aids seem to perform better with the new hearing aids, and equally well for both programs.

The normalised *gain* (G_E) was used to compare the average gains between the three conditions. Note that $G_E = 0$ when the *gains* for two conditions are the same (see 'Patients and methods'). Figure 5 demonstrates that the overall mean values did not differ significantly.

Overall, on all parameters, in all conditions of the sound localisation test, no significant differences in sound localisation were found between the own and new hearing aids nor between the two programs in the new hearing aids. For the average values of these parameters, see Table S1 (A and B).

Questionnaires

Figure 6 shows the results of the AHPAB per subscale compared to the norm percentiles obtained in 2010 by Johnson *et al.*[28] for successful hearing aid users. When comparing the AHPAB concerning the previous hearing aids and that concerning the new hearing aids, on each subscale a decline in disability was seen, which was statistically significant on the subscale ease of communication ($P = 0.018$; 95% CI 2.47–22.69). All items in the subscale ease of communication concerned situations in quiet, in small groups without background noise. Furthermore, the overall score on the APHAB improved significantly (from 46.0, SD 12.4 to 34.8, SD 12.5, $P = 0.018$; 95% CI 2.02–18.46), representing an overall benefit of the new hearing aids compared to their own hearing aids. Concerning listening in background noise or in reverberant places, no significant changes were found.

On the GHADP questionnaire, a high score on device use is reported, with median scores above 90% for their own as well as new hearing aids. After using the new hearing aids, the mean reported disability, in pre-determined and personal relevant situations, showed a significant drop of 16.3% on a baseline of 58.6% ($P = 0.002$).

Finally, using their diary, the patients reported a major decrease in change of programs between the first and last week of the study, from 6.6 (SD 4.6) to 2.4 (SD 1.7) changes per day ($P = 0.001$). More important, the satisfaction with the linear program, 7.7 (range 1.0–9.6), was significantly higher than the satisfaction with the nonlinear program, which was 5.9 (range 1.1–8.5) ($P = 0.02$).

Discussion

The present study showed that the included patients with Usher syndrome type IIa demonstrate a significant preference for the linear amplification program with

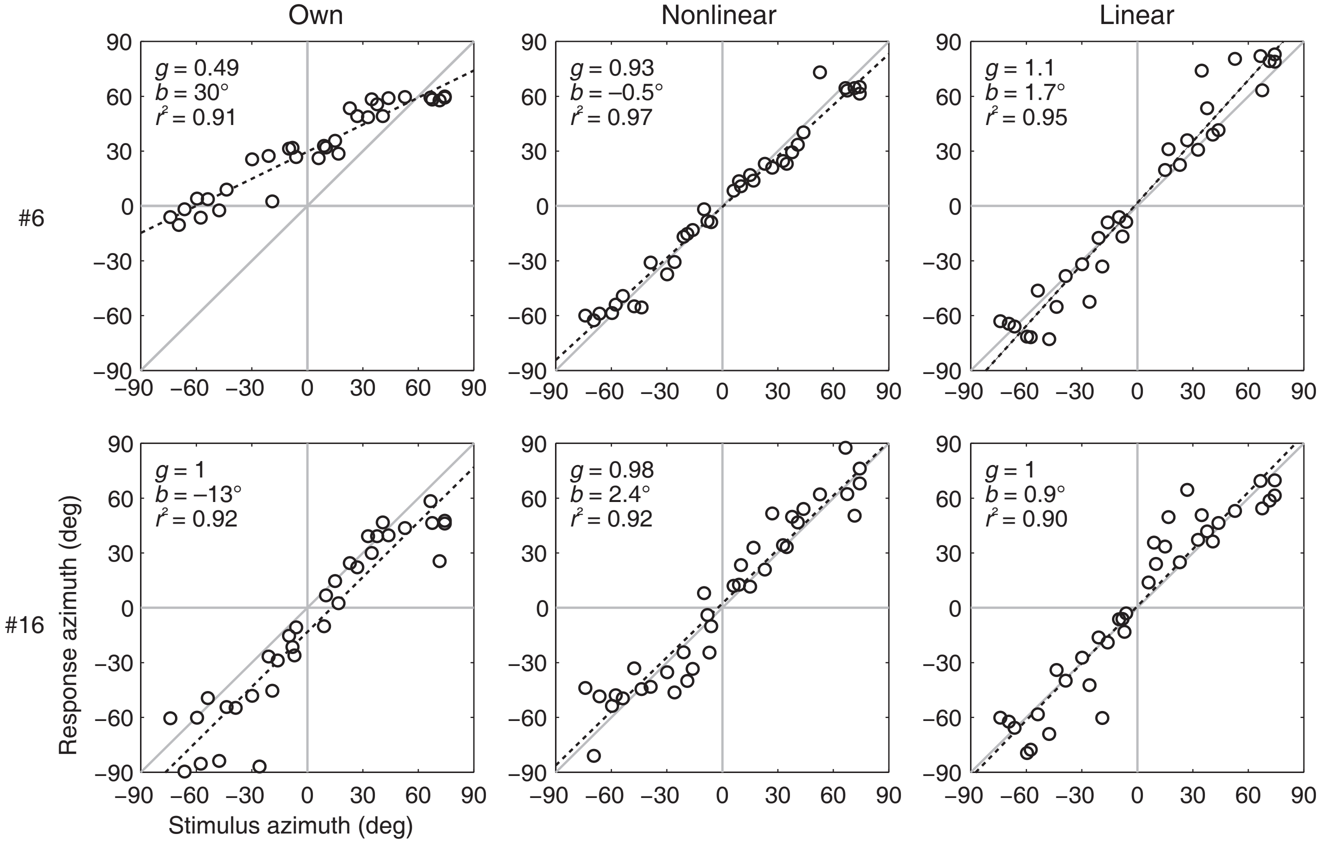

Fig. 3. Sound localisation in azimuth in three conditions for two patients. Graph representing the results of two individual patients (#6 and #16) for sound localisation in azimuth (horizontal plane) in three conditions: with their own hearing aids, with the new hearing aids with the nonlinear amplification program and with the new hearing aids with the linear amplification program. Each dot represents one of the 36 broadband stimuli. The dotted line represents the best linear fit (least squares criterion) of the stimulus–response relationship. The parameters of the fit are shown in the panel: g = response *gain*, b = response *bias*, and r^2 = coefficient of determination (see 'Patients and methods'). Abbreviations: deg, degrees.

omnidirectional microphone over the nonlinear program with pinna imitating directionality, measured objectively (mean logged use of 77% *versus* 17%) as well as subjectively (satisfaction of 7.7 *versus* 5.9 out of 10). In addition, the self-reported outcomes showed significantly higher scores on ease of communication and overall benefit and lower scores on disability for the new hearing aids compared to their own hearing aids. These results are complemented with good results for speech intelligibility in noise and sound localisation. The latter tests, however, did neither show a difference between hearing aids nor programs. It should be noted that the two domains of the APHAB that deal with spatial hearing (hearing in noisy places and in reverberant surroundings) did not show a significant improvement, which seems to be in agreement with the objective measurements. Therefore, the hypothesis that a linear amplification program of hearing aids in Usher syndrome type IIa patients would lead to improved sound localisation when compared to a nonlinear amplification setting could so far not be confirmed.

On an individual level, differences in localisation were found between the patient's own hearing aids and their new hearing aids at follow-up for BB (Fig. 5) and LP sounds. Patients who performed poorly with their own hearing aids performed better with the new hearing aids in either program. Patients who already performed quite well with their own hearing aids, performed equally well with the new hearing aids in both programs.

The good sound localisation performance of the patients with Usher syndrome type IIa is not in agreement with their reported subjective difficulties.[10] A possible explanation for these differences might lie in the validity of the used method. The laboratory set-up and test protocol used in this study may not appropriately assess the reported difficulties.

Further research is needed to evaluate the experienced difficulties by these patients. Possible interfering difficulties

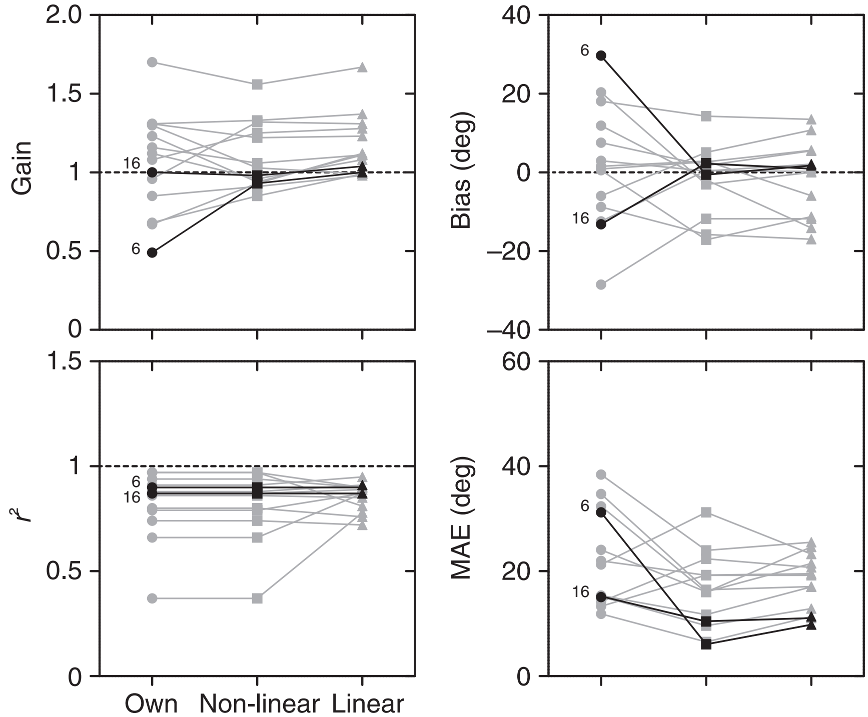

Fig. 4. Individual sound localisation parameters. Graphs representing individual *gain*, bias, coefficient of determination and Mean Absolute Error (MAE) values for broadband stimuli in three conditions: with the patient's own (dots) and new hearing aids in nonlinear (squares) and linear (triangles) program. The values for patients #6 and #16 are highlighted for they were represented in Fig. 3.

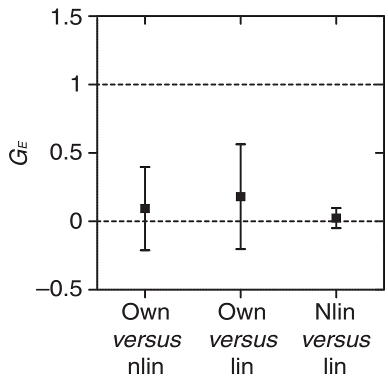

Fig. 5. Comparison of normalised average gains. Graph representing the mean *gain*-error (G_E) for the differences between the own and new hearing aids in the nonlinear amplification program (nlin), between the own and new hearing aids in the linear amplification program (lin) and between the nonlinear amplification program of the new hearing aids. Per mean G_E, the standard deviations are represented by black lines on either sides of the squares.

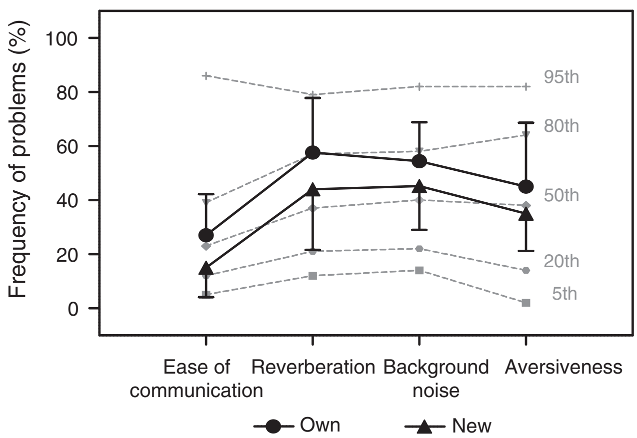

Fig. 6. Abbreviated Profile of Hearing Aid Benefit (APHAB). Mean scores for the AHPAB subscales for the own (black squares −SD) and new (black triangles +SD) hearing aids. For comparison, the norm percentiles as defined by Johnson *et al.* in 2010 for successful hearing aid users were added.[28]

might be distance estimation, moving sound stimuli, localisation of sound stimuli in noise or reverberation.

In 2006, Keidser *et al.* performed localisation tests on hearing impaired patients with different compression techniques and directional microphone settings. They could not detect any difference in left–right localisation between a linear program with omnidirectional microphone and a program with syllabic compression and omnidirectional microphone. These results corroborate the findings of the present study. However, the nonlinear program (with syllabic compression) in our study was complemented by a moderate static directional microphone. A possible explanation for the absence of a difference may lie in the localisation environment. In our study, no noise was presented during the localisation task. A more complex localisation task, with noise, might accentuate any differences in program or microphone setting.

In conclusion, the examined patients with Usher syndrome type IIa prefer the linear program over the nonlinear program. However, apart from a significantly higher logged use, no difference in speech in noise and sound localisation was observed. Further research is needed to address the preference of a linear over a nonlinear amplification program and to replicate the present results in hearing impaired patients without additional visual impairment.

Keypoints

- Patients with Usher syndrome type IIa need adequate hearing aid fitting due to their double-sensory impairment.
- Patients with Usher syndrome type IIa prefer a linear over a nonlinear hearing aid program.
- No difference in speech in noise and sound localisation was observed between the linear and nonlinear hearing aid program.

Conflict of interest

None to declare.

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