


## CASE REPORT

# Fluctuation in electrical hearing in a Morbus Meniere's patient

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**Abstract**

The glycerol test is an easy-to-use instrument to elucidate fluctuations of electrical hearing in patients with Meniere's disease and it might be also used as a therapeutic option.

**KEYWORDS**

cochlear implant, meniere's disease, hearing fluctuation, ECAP, glycerol test

## 1 | INTRODUCTION

Meniere's disease (MD) is a disorder of the inner ear, characterized by episodes of vertigo, tinnitus, and hearing loss. The mechanism of its development is still unclear, but raised endolymphatic pressure is commonly accepted as a major cause.

Cochlear implantation is the routine treatment for severe hearing loss in patients with MD. Speech perception outcomes with cochlear implant (CI) vary among patients with MD, but a significant improvement in word and sentence tests similar to the overall CI population is described.<sup>1,2</sup> Although there is a high residual low-frequency hearing preservation rate after implantation, the long-term hearing preservation for these patients is even worse than in non-MD patients.<sup>3</sup> It is suggested that the baseline hydrops is responsible for the long-time hearing loss of the residual hearing.

There is also evidence for the fluctuation in electrical hearing after cochlear implantation in patients with MD: McNeill and Eykamp<sup>4</sup> reported a case of high fluctuations in the electrical impedance of a patient with MD implanted with a cochlear implant and distortion in speech perception, where simple reprogramming of the CI system was able to solve these problems.

The underlying mechanism for the fluctuation in hearing in patients with MD is still speculative: The endolymphatic

hydrops might alter the electrode position or disturb the electrode-neural interaction by changing the endolymph fluid potential.

Herein, we describe a case where the glycerol test, introduced by Klockhoff and Lindblom in 1967<sup>5</sup> to detect MD, improved the electrical hearing in an acute period of hearing loss of a patient with MD. Glycerol, given perorally as a single dose, produces a transient hyperosmosis in the blood and lowers the intralabyrinthine pressure. As a consequence, the effect of the endolymphatic hydrops is lowered within a short time period. In our case, an improvement of electrical hearing thresholds was measurable 2 hours after the intake of glycerol. Electrically evoked compound action potentials (ECAPs) were used to monitor cochlear nerve responses.

## 2 | CASE REPORT

A 48-year-old female patient suffering from bilateral MD for 15 years and significant progredient hearing loss, wearing bilateral hearing aids for 2 years, complained about worsening of her speech understanding and fluctuations in hearing. After the CI evaluation, she underwent cochlear implantation on the inferior side in September 2019. She was also suffering from tinnitus on both ears (250 Hz noise, visual analogue

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scale 95 of 100) and a glycogenosis type IX affecting the liver phosphorylase b kinase.

The CI surgery was a routine round window insertion monitored with flat-detector computed tomography showing a regular inserted electrode array (CI632, Cochlear™). Regular intraoperative ECAP thresholds were recorded at all 22 intracochlear electrodes.

Within 6 weeks after the first fitting, her hearing ameliorated from 15% monosyllabic perception at 65 dB SPL in a free field condition with hearing aid to 80% with CI (CP1000 speech processor). Aided hearing thresholds with CI were between 30 dB and 15 dB for frequencies between 250 and 6000 Hz. She did not mention any fluctuations in electrical hearing.

Five months after implantation, the patient reported a massive degradation of understanding with the speech processor for about 10 days and a hollow sound without any other symptoms such as vertigo or worsened tinnitus. The technical control showed a regular working sound processor with intact microphones and unchanged regular electrical impedances of the electrodes.

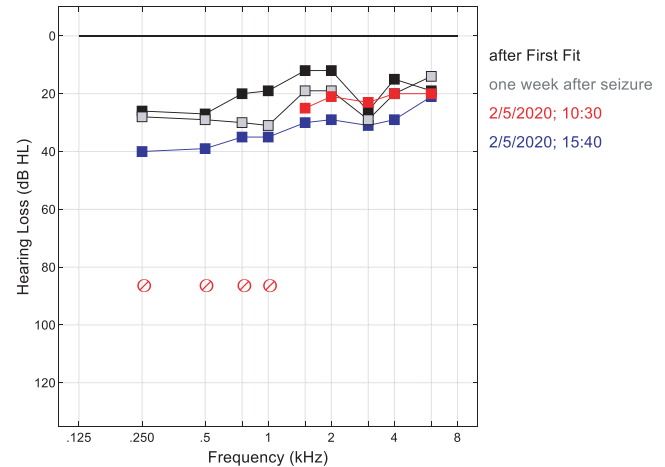
Aided thresholds were not measurable up to 1000 Hz, and 20 dB HL for frequencies at 1500 Hz and higher; monosyllabic understanding was 0%. ECAPs measured via AutoNRT™ showed no responses for the apical electrodes (el. 11-22) representing frequencies below 2063 Hz and nearly unchanged responses in the basal electrodes (el. 1-8) representing frequencies above 2688 Hz. Electrical stimulation of the apical electrodes 11-22 corresponding to frequencies 188-2063 Hz did not result in any hearing.

To verify that an episode of Meniere active disease was responsible for this deterioration in hearing, the Klockhoff test<sup>5</sup> was performed. Glycerol was given perorally as a single dose of 1.5 g/kg body weight, and the effect on the residual hearing was monitored by repeated pure tone audiometry every 30 minutes. Electrical hearing with the speech processor was determined 2 hours after the intake of the glycerol solution. One week after the episode, aided thresholds with the CI were determined.

Impedances, ECAP thresholds, and pure tone audiometry were again measured during a follow-up visit 6 weeks later.

While the glycerol test showed no significant effect on the residual hearing, audiometric thresholds measured with the speech processor recovered. Aided thresholds were between 35 and 40 dB for 250-1000 Hz and 20 to 30 dB for frequencies at 1500 Hz and higher. Accordingly, the patient recognized a better speech understanding. Two hours after the glycerol application the patient regained a 60% speech understanding of monosyllables.

One week after the episode, aided thresholds were around 35 dB for 250-1000 Hz and 15 to 30 dB for frequencies at 1500 Hz and higher (Figure 1).



**FIGURE 1** Free field audiometry of the electrical hearing before and after treatment with glycerol; squares symbolize measured hearing levels and slashed zeros denote no response within the limits of the audiometer. The red curve was measured before the treatment with glycerol, and the blue curve was measured 2 h after the intake

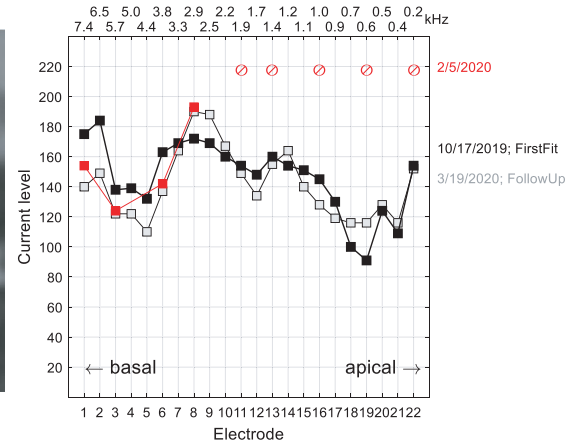
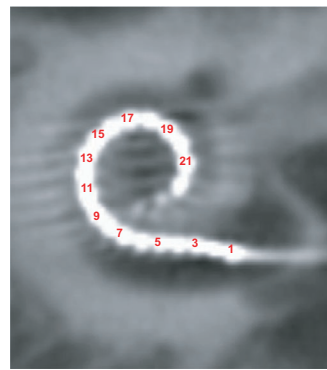
During the follow-up visit 6 weeks later, the patient reported a good speech understanding with the CI. ECAPs were comparable to the FirstFit data (Figure 2), and the monosyllabic understanding was 90%.

We measured impedances of the implant pre- and post-treatment with glycerol, and there were only marginal fluctuations measurable, not correlated with the sudden hearing loss (Figure 3).

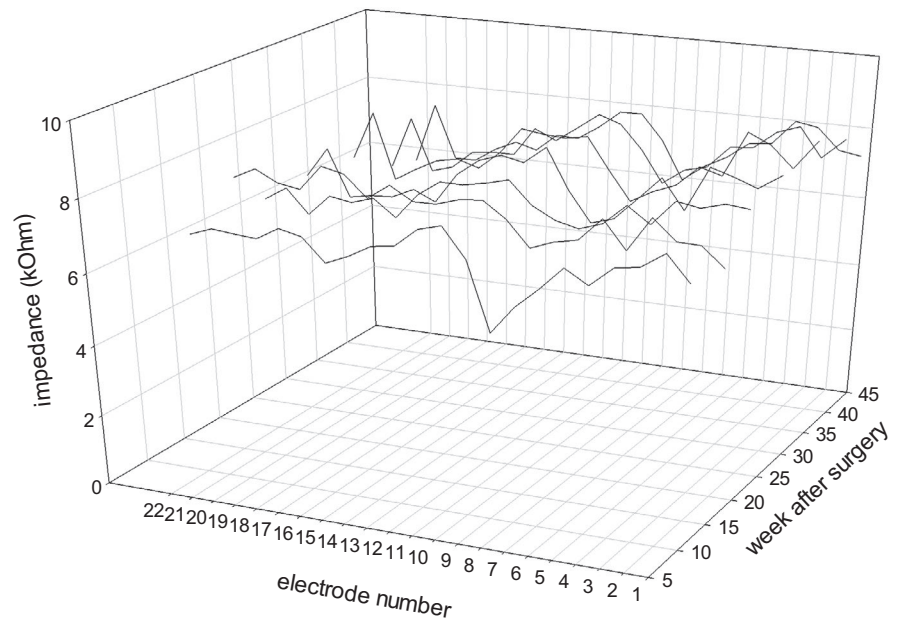
### 3 | DISCUSSION

Fluctuation in hearing is a known problem for patients with MD and a challenge in the assessment of CI candidacy. The symptoms of our patient showed for the first time that this fluctuation is also present in MD patients with CI, affecting not only the residual hearing, but also the electrical hearing, measured by ECAPs and aided pure tone audiometry. It has been hypothesized that during a crisis of Meniere active disease, the intracochlear position of the electrodes is altered. McNeill and Eykamp<sup>4</sup> assume that electrical resistance changes of the blood-labyrinth barrier create fluctuations in the CI performance, but the detailed mechanism underlying this remains unclear. These changes seem to be reversible by the intake of an osmotic diuretic drug, which dehydrates the inner ear and reduces the inner ear pressure back to normal. In our case, the effect of the glycerol test was permanent for the observed 6-week follow-up; the patient reported a stable good speech understanding. We cannot exclude that further episodes of Meniere active disease will again alter the electrical hearing in this patient, and we are not sure whether the hearing would have ameliorated accordingly without the glycerol treatment. Compared with

**FIGURE 2** Left: CT scan after surgery. Right: ECAP thresholds at CI activation (black), on the day of the reported hearing loss (red) and 6 wk later (gray). Squares symbolize measured ECAP thresholds, and slashed zeros denote no valid ECAP response



**FIGURE 3** Impedance measurements over time of the implanted electrode (CI632, Cochlear™)



3T magnetic resonance imaging after intratympanic injection of contrast agent, the glycerol test has a sensitivity of only 55%,<sup>6</sup> but is more easier to implement and may be a therapeutic option.

## 4 | CONCLUSION

It is important to counsel patients with MD before surgery, that hearing fluctuations may also appear with electrical hearing. The glycerol test might be an easy-to-use instrument to uncover this fluctuation of hearing and may serve as a therapeutic option.

## ACKNOWLEDGMENT

Published with written consent of the patient.

## CONFLICT OF INTEREST

None declared.

## AUTHOR CONTRIBUTIONS

AH: completed the background research and drafted and edited the manuscript. ME, JH: involved in the patient care and performed tests. TL: edited the manuscript and involved in figure development. UH: edited the manuscript and approved the final manuscript.

## ETHICAL APPROVAL

Ethical approval (162\_17Bc) has been given by the IRB of the Friedrich-Alexander-Universität Erlangen-Nürnberg. The patient provided written informed consent for the publication.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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