

Auditory steady state response in auditory neuropathy

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Dear Sirs,

We read with interest Emara and Gabr's article¹ promoting auditory steady-state response assessment as a physiological test of hearing threshold, as part of the test battery for patients with auditory neuropathy spectrum disorder. The limited utility of auditory brainstem response (ABR) testing in predicting behavioural thresholds in patients with this disorder is a significant weakness in the current diagnostic test battery used to quantify the degree of hearing loss in such infants, and the lack of a physiological test to estimate behavioural thresholds may cause delays in intervention. Currently, clinicians must wait until behavioural thresholds can be determined reliably before making informed decisions concerning amplification or cochlear implantation. In their article, Emara and Gabr conclude that auditory steady-state response assessment may provide a valid alternative to behavioural threshold estimation, and they recommend its use to complete the evaluation of patients with this disorder.

Despite the need for an objective assessment, we have many concerns with Emara and Gabr's conclusions and recommendations. These authors suggest that the auditory steady-state response assessment is preferable to ABR assessment, primarily on the basis that the auditory steady-state response was measured in 10 out of 13 participants who had the disorder, whereas the ABR was absent in all but two of the cases. However, the measured auditory steady-state response thresholds were not significantly correlated with measured behavioural thresholds at any frequency tested. This lack of correlation, and the lack of raw data or descriptive statistics, leaves the reader unable to determine the range of clinical error that could potentially be made by using thresholds obtained by auditory steady-state response measurement when assessing the need for intervention. The varying direction and wide range of non-significant correlations between auditory steady-state response threshold and behavioural thresholds across frequencies (−0.529 to +0.732) would also preclude accurate estimation of the degree and configuration of the hearing loss.

Previous studies have reported no correlation between auditory steady-state response threshold and behavioural threshold for these patients,^{2–4} and have reported that auditory steady-state response thresholds range from 20 to 70 dB or more around the behavioural

thresholds for this population. With such variability, a patient with a moderate hearing loss as determined by auditory steady-state response assessment could potentially have a normal behavioural threshold or a severe hearing loss, or any degree of loss between these two extremes. This lack of a predictable relationship between auditory steady-state response and behavioural threshold limits the usefulness of the test.

In addition, Emara and Gabr do not discuss previous literature citing the presence of measurable auditory steady-state responses at levels lower than the listener's behavioural threshold. Gorga and colleagues⁵ measured auditory steady-state responses in 10 patients with profound hearing loss, and found levels that were 20 dB less on average than the subjects' behavioural thresholds. In some cases, auditory steady state responses were measured in ears with no behavioural response to sound. Such findings have led to the conclusion that auditory steady-state responses recorded at high intensity levels may be related to artefacts, rather than being true reflections of peripheral auditory function. While other studies have suggested strategies to minimise such artefacts,⁶ omission of this consideration from Emara and Gabr's discussion makes it difficult to conclude whether their investigation took such factors into account, or whether this could be an underlying factor in the significant variability observed in the auditory steady-state response data.

A limitation of auditory steady-state response testing is that two important diagnostic characteristics of auditory neuropathy spectrum disorder may be obscured, as the measurements use Fourier analyses in the time domain. The absent ABR response does not provide an estimate of the behavioural threshold, but is in itself an important characteristic differentiating auditory neuropathy spectrum disorder from other types of hearing loss. Given that auditory steady-state response thresholds are not correlated with behavioural thresholds in this population, the ability to measure auditory steady-state response thresholds could make diagnosis of auditory neuropathy spectrum disorder more difficult, particularly in cases where such thresholds are normal or minimally depressed. In auditory neuropathy spectrum disorder patients, cochlear microphonic parameters may indicate the functionality of the cochlear outer hair cells. The auditory steady-state response does not assess these parameters; however, assessment of otoacoustic emissions together with auditory steady-state response may help to characterise outer hair cell function (although previous studies have indicated that otoacoustic emissions are

not always present, and can also disappear over time, recording the cochlear microphonic (CM) may indicate normally functioning cochlear outer hair cells (OHC) in these patients, and is not recorded in the auditory steady-state response, and otoacoustic emission (OAE) testing with auditory steady-state response could help to characterize OHC function, although previous studies have indicated that otoacoustic emissions are not always present or can disappear over time in these patients).

Emara and Gabr recommend using auditory steady-state response measurement in conjunction with ABR and otoacoustic emission testing to complete the diagnostic evaluation of such patients. However, given the poor correlation between auditory steady-state response thresholds and behavioural thresholds, there appears to be no clinical utility in adding auditory steady-state response measurement to the test battery used to assess patients with auditory neuropathy spectrum disorder.

In summary, we remain concerned that some clinicians may potentially be misled by the assertion that auditory steady-state response testing is a valid means of predicting auditory sensitivity in patients with auditory neuropathy spectrum disorder.

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Author's reply

Dear Sirs,

We thank Drs McCreery and Simmons for their comments regarding our paper on auditory steady-state response testing in auditory neuropathy. We used two types of evoked potentials, auditory brainstem response (ABR) and auditory steady-state response, and the aim of our work was to test the utility of auditory steady-

state response (ASSR) testing in the assessment of hearing thresholds. An auditory steady-state response was recorded in 10 of the 13 cases studied; for these cases, auditory steady-state response thresholds were higher than behavioural thresholds at all frequencies. We mentioned the mean difference and confidence interval between both thresholds, and our work indicated that auditory steady-state response thresholds were worse than behavioural thresholds in patients with auditory neuropathy. In such patients, a measurable auditory steady-state response threshold probably indicates a better behavioural threshold. The auditory steady-state response can therefore provide a useful, although not entirely accurate, indication of hearing threshold and configuration. Caution is obviously required in patients with an absent auditory steady-state response.

Drs McCreery and Simmons mention the work of Gorga *et al.*,¹ who measured auditory steady-state responses in 10 patients with profound hearing loss, and found levels that were 20 dB less on average than the patients' behavioural thresholds. In our work, we commenced auditory steady state response measurement at 90 dB; when a response was obtained, the intensity was lowered in 10 dB steps until the threshold was reached. Our equipment was capable of delivering sound stimuli of up to 118 dB; however, we did not increase the intensity above 90 dB to avoid aliasing and artefact, as well as non-auditory responses to high intensity stimuli. We also used monaural, single frequency recordings to avoid frequency interaction, which may affect auditory steady-state response measurement.

It is widely known that the diagnosis of auditory neuropathy depends upon certain criteria: normal otoacoustic emissions and/or normal cochlear microphonics, together with absent or severely abnormal ABRs that are out of proportion with the pure tone audiogram. Auditory brainstem response testing is clearly the cornerstone of auditory neuropathy diagnosis, and cannot be replaced by another test. In this regard, we do not suggest that auditory steady-state response testing can replace ABR testing. However, we do believe that auditory steady-state response testing can be used alongside other tests as a useful adjunct to the evaluation of ABR in patients with auditory neuropathy.

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