BIAP Recommendation 06/17:

Hearing aid fitting in children with Auditory Neuropathy Spectrum Disorder (ANSD)

General foreword

This document presents a Recommendation by the International Bureau for Audiophonology BIAP.

A BIAP Recommendation provides a reference standard for conducting an audiological or phonological intervention that represents, to the best knowledge of BIAP, the evidence base and good practice concerning the stated methodology and scope of the document at the time of publication.

Although care has been taken in preparing the information supplied, BIAP does not and cannot guarantee its interpretation or application. BIAP cannot be held liable for any errors or omissions, and BIAP accepts no liability whatsoever for any loss or damage howsoever arising. This document shall be effective until superseded or withdrawn by BIAP.

Comments on this document are welcome and should be sent to the Secretary General of the International Bureau for Audiophonology BIAP. The address can be found on the BIAP website at www.biap.org.

Introduction

An Auditory Neuropathy Spectrum Disorder (ANSD) imposes the need for reflection and deep consideration since there are still many uncertainties about its clinical treatment. For this reason, many audiologists feel uncomfortable when fitting hearing aids in children with ANSD. In the past, there were contradictory proposals about the use of hearing aids (HAs) in this population.

A recommendation on the assessment of ANSD is available in REC TC12-09.

There is increasing evidence that the HA-fitting process must be started as soon as reliable behavioural responses are available, in order to stimulate the auditory pathway in time. Several studies have shown that children with ANSD can benefit from HA and/or cochlear implants (CI) (Ching et al, 2013; Fitzpatrick et al, 2019). At the same time, it will be important to look for the aetiology and localisation of the ANSD as it will influence clinical decision-making.

Scope

This recommendation will focus especially on the selection and fitting of hearing aids in young children with ANSD. An interdisciplinary approach in partnership with the parents is required in this population.

Recommendation

1. Audiological assessment

   In cases of ANSD, it is necessary to obtain the best estimation of the threshold based on minimal response levels with **behavioural testing** with bone- and ear-specific air conduction, since ABR is not a predictor of hearing thresholds in this population

(Rec 12/08). Only behavioural thresholds can be used for determining gain and output of the hearing aid.

In case of fluctuating thresholds, it will be necessary to rule out middle ear ventilation problems, and to use the lowest established thresholds to avoid over-amplification.

Cortical auditory evoked potentials or **CAEP-testing** in awake infants can give some useful clinical information of cortical auditory maturation or behavioural performance. P1 CAEP responses can provide valuable information regarding the maturational status of the auditory cortex. But there is still some controversy in the clinical use of this tool in young hearing-impaired children (British Society of Audiology [BSA], 2019).

2. Selection and fitting of hearing aids

The American Academy of Audiology (AAA) Guidelines on paediatric amplification (2013) recommend that children with ANSD always have a trial with hearing aids if auditory thresholds are insufficient to support speech perception at conversational levels, regardless the degree of hearing loss.

Children with ANSD have often greater temporal processing deficits and reduced frequency discrimination, which can be associated with poorer speech perception in quietness but also, and especially, in noise.

The main focus on the selection of hearing-aid features will be the **speech in noise management**. Other features such as noise reduction, directional systems (multi-directional microphones), binaurally-linked microphones, automatic multiple programmes…will be required (Scollie & Bagatto, 2020).

Higher technology to maximise audibility and minimise distortion of the input signal will be needed.

In addition, connectivity with assistive listening devices should always be available.

The initial fitting of the hearing aids should be based on a prescriptive method for children (DSL V5 or NAL NL2) that uses behavioural responses. It is important to keep in mind that the behavioural thresholds still may include some uncertainties and must be re-evaluated regularly. Changes in the hearing thresholds require adjustments in the fitting of the hearing aid.

A previous study by Prabhu & Barman (2017) with older children with a moderate hearing loss suggest the reduction of the amplification in low frequencies for individuals with ANSD to avoid the upward spread of masking. Other studies with an adult population with ANSD suggest that hearing aids with slow compression time settings or linear amplification may be chosen over those with fast compression as they allow greater amplitude differences in the temporal modulation, providing better speech quality (Mathai & Appu, 2015; Spirakis, 2011). At this moment, with the new technology, there are more options to choose from and to combine from the available algorithms (fast or slow compression, polarity of microphones, noise management…) that may make it more efficient for children with ANSD.

3. Verification and validation

Electro-acoustic measurements and **behavioural testing** in combination with clinical observation must be done carefully and on a regular basis.

Optimal audibility of speech must be verified with real ear or testbox measurements. The Speech Intelligibility Index (SII) must be evaluated with the aided SII normative data form. The adjustment should always be fit to target. Normative data are well described by Moodie at al (2017)[1].

Behavioural testing should focus on auditory speech perception skills in quietness and in noise. An auditory speech sound evaluation (discrimination of phonemes) is important for evaluating spectral cues (Like A§E®). Especially in young children, where speech audiometry is not yet possible, the use of speech sound evaluation is recommended.

Questionnaires can help the evaluation of the functional outcomes of the hearing aid fitting and the use of Hearing Assistive Technology (HATs). Helpful questionnaires are: LittlEARS auditory questionnaire, IT-MAIS (Infant Toddler Meaningful Auditory Integration Scale), PEACH (Parent Evaluation of Aural/Oral Performance of Children), TEACH (Teacher's Evaluation of Aural/Oral Performance of Children), ELF (Early Listening Function), CAP (Categories of Auditory Performance), FAPI (Functional Auditory Performance Indicators).

Scientific evidence suggests the **CAEP-test** is proven to be useful for clinicians regarding the follow up and rehabilitation of these patients. (Sharma at al, 2011). Especially those children with lack of improvement of auditory skills should have their long latency potentials monitored over time to ensure that the potential benefit of cochlear implants is chosen as an alternative before the deprivation effect obstructs a further rehabilitation process. However, CAEP-measurements are not yet widely used.

According to Cardon & Sharma (2013), the sensitive period for intervention of children with ANSD that receive CIs is around 2 years of age.

A close interaction among professionals dealing with hearing aid evaluation and fitting, and the rehabilitation process, is mandatory to guarantee the maximum benefit from the intervention process.

4. Follow up

All children with ANSD must be enrolled in an **early intervention programme** including aural rehabilitation and supplemental visual communication (cues) if needed.

Children with ANSD may have extreme difficulties in understanding speech in noise. Therefore, a trial of **personal HATs** must be part of the fitting process (Rec 06/16-07/07).

Data logging can help to monitor daily use and have an insight in the acoustical environment.

An ongoing evaluation of hearing, speech and language development, and of the communication skills, is essential.

5. Decision CI or HA

When no progress on the development of speech, language, and communication with conventional hearing aids is seen, a cochlear implant can be an option. This process takes more time than with children with a sensorineural hearing loss without ANSD (Rec 12/09).

The decision for the CI must not be based upon the electrophysiological results, but on stable behavioural responses. A cochlear implant should be a final alternative only after

audiological findings are reliable and confirm permanent ANSD. Nonetheless, these children need constant support for language development and communication skills.

[1] https://www.dslio.com/wp-content/uploads/2016/10/Aided-SII-Normative-Values_App-A.pdf

References

American Academy of Audiology [AAA]. (2013). *American Academy of Audiology Clinical Practical Guidelines Pediatric Amplification*. Retrieved from: http://galster.net/wp-content/uploads/2013/07/AAA-2013-Pediatric-Amp-Guidelines.pdf

British Society of Audiology [BSA]. (2019). Recommended Procedure: *Assessment and management of Auditory Neuropathy Spectrum Disorder (ANSD) in Young Infants*. Retrieved from: https://www.thebsa.org.uk/wp-content/uploads/2019/01/FINAL-JAN2019_Recommended-Procedure-Assessment-and-Management-of-ANSD-in-Young-Infants-GL22-01-19.pdf

Cardon, G., & Sharma, A. (2013). Central auditory maturation and behavioral outcome in children with auditory neuropathy spectrum disorder who use cochlear implants. *International Journal of Audiology, 52*(9), 577-586. https://doi.org/10.3109/14992027.2013.799786

Ching, T.C., Day, J., Dillon, H., Gardner-Berry, K., Hou, S., Seeto, M., Wong, A., Zhang, V. (2013). Impact of the presence of auditory neuropathy spectrum disorder (ANSD) on outcomes of children at three years of age. *International Journal of Audiology*, *52*: S55–S64. https://doi:10.3109/14992027.2013.796532

Fitzpatrick, E.M., Cologrosso, E., & Sikora, L. (2019). Candidacy for Amplification in Children With Hearing Loss: A Review of Guidelines and Recommendations. *American Journal of Audiology 28*(4). https://doi.org/10.1044/2019_AJA-19-006

Gabr, T.A. (2016) Amplification Options in Children with Auditory Neuropathy Spectrum Disorder. *Communication Disorders, Deaf Studies & Hearing Aids 4*(1). https://doi:10.4172/2375-4427.1000151

The Joint Committee on Infant Hearing. (2019) Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *The Journal of Early Hearing Detection and Intervention 4*(2):1-44. https://doi:10.15142

Mathai, J. & Appu, S. (2015) Perception of hearing aid–processed speech in individuals with late-onset auditory neuropathy spectrum disorder. *Journal of the American Academy of Audiology 27*(2). https://doi:10.3766/jaaa.14102

McCreery, R.W., & Walker E.A. (2017). *Pediatric Amplification: Enhancing Auditory Access.* Plural Publishing, San Diego.

Moodie, S.T.F., The Network of Pediatric Audiologists of Canada, Scollie, S.D., Bagatto, M.P., & Keene Kelly. (2017). Fit-to-Targets for the Desired Sensation Level Version 5.0a Hearing Aid Prescription Method for Children. *American Journal of Audiology 26*(3),251-258. https://doi.org/10.1044/2017_AJA-16-0054

Prabhu, P. & Barman, A. (2017). Effectiveness of Low-Cut Modified Amplification using Receiver in the Canal Hearing Aid in Individuals with Auditory Neuropathy Spectrum Disorder. *International Archives in Otorhinolaryngology 21*(3):243–249. https://doi.org/10.1055/s-0036-1593471

Roush, P. (2020). Clinical Management of ANSD in Infants and Young Children. (2020). *Canadian Audiologist 7*(5). https://canadianaudiologist.ca/issue/volume-7-issue-3-2020/pediatric-audiologist-feature/

Scollie, S. & Bagatto, M. (2020) Noise Management in Pediatric Hearing Aid Fitting. *Canadian Audiologist 7*(5). https://canadianaudiologist.ca/issue/volume-7-issue-3-2020/noise-management-pediatric-feature/

Sharma, A., Cardon, G., Henion, K., & Roland, P. (2011). Cortical maturation and Behavioral outcomes in children with auditory neuropathy spectrum disorder. *International Journal of Audiology 50*(2), 98–106. https://10.3109/14992027.2010.542492

Spirakis S. (2011) Auditory Neuropathy Spectrum Disorder and Hearing Aids: Rethinking Fitting Strategies *Hearing Review*. 2011;18(11): 28-33.

Walker E, McCreery R, Spratford M, and Roush P. (2016) Children with auditory neuropathy spectrum disorder fitted with hearing aids applying the American Academy of Audiology Pediatric Amplification Guideline: Current Practice and Outcomes. J Am Acad Audiol 2016;27(3):204–18. https://doi:10.3766/jaaa.15050. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4789798/

This recommendation was created and approved in a multidisciplinary cooperation between professionals of all audiophonologic disciplines, which are medicine, pedagogy, speech therapy, psychology, and hearing instrument audiology.

The original language of this document is English.

President of the commission TC06: Kristin Kerkhofs (Belgium)

Members of the commission TC 06: E. Boechat (Brazil), A. Bohnert (Germany), M. Chapchap (Brazil), A. Coez (France), S. Cozma (Romania), L. Demanez (Belgium), M. Maggio (Spain), A. Jouarez (Spain, A. Kerouedan (France), V. Klinck (Belgium), G. Madeira (Belgium), E. Orzan (Italy), S. Ramlow (Germany), C. Renard (France), H.Thai-Van (France), K. Tiede (Germany), C. Vander Heyden (Belgium), P. Verheyden (Belgium), Th. Wiesner (Germany), F. Zajicek (Austria).

Zoom Meeting, May 1st, 2021

Keywords: Auditory Neuropathy Spectrum Disorder, ANSD, hearing aid fitting, infant, assessment.