International Journal of Brain and Cognitive Sciences 2013, 2(2): 9-13
DOI: 10.5923/j.ijbcs.20130202.01

Study of Audiological Features, Problabe Etiology and Neurological Aspects of Children with Auditory Neuropathy/Hearing Dyssinchrony

Roney W. D. de Oliveira[1], Bianca P. Lanzetta[2], Maria Bernadeth de S. Freitas[2], Kathia R. X. Vale[2], Luziene D. B. de Oliveira[2,*]

[1]Department of Phisiology, UFES, Vitória, Espirito Santo, Brazil
[2]Department of Audiology, Policlinica de Referência UVV, Vila Velha, Espirito Santo, Brazil

Abstract The auditory neuropathy is a nosologic entity characterized by the loss of the synchronous auditory nerve activity associated with the preservation of cochlear function. Objectives: To describe auditory features, probable etiologies, and neurological aspects of children with auditory neuropathy from an Auditory Health Program. Method: Retrospective analysis of the children's records seen in an Auditory Health Program of the Polyclinic of UVV, from December 2005 to December 2010. We selected 12 children with suspected auditory neuropathy as presenting cochlear integrity, confirmed by the presence of otoacoustic emissions, tympanometry preferably type A, and alterations of the rest of auditory canal, suspected by the presence of the cochlear microphonic in the Brainstem Auditory Evoked Potential (BAEP) with varying degree loss, and behavioral audiometry or tonal audiometry were better than the expected based on the results of BAEP. Farther, we revised the neurologic anamnesis. Results: The BAEP found no response with the presence of cochlear microphonic (CM) in 100% of cases. We also noted predominance of transient otoacoustic emission absent, cochlear-Eyelid Reflex, and estapedic reflex absent. Incompatibility of responses was confirmed among the tests when compared to the psychoacoustic and auditory behavior. Concerning the etiological factors and clinical history, only four children had no neonatal complications that could explain auditory findings, three other patients had prematurity, hypoxia (3 cases), bilirubin encephalopathy (1 case) or sepsis (5 cases) with the use of ototoxic drugs in the neonatal period. Conclusion: The auditory assessment shows that the findings are characterized by variability of response in the tonal audiometry with inconsistent responses, and a development of listening skills below the chronological age. Thus, we conclude that the tests should never be analyzed isolated. Besides that, it's important to listen attentively to the complaints of the parents and caregivers.

Keywords Auditory Neuropathy / Hearing Dyssinchrony, Audiological Features in Children, Cochlear microphonic, Health Hearing

1. Introduction

Auditory neuropathy (AN) is a nosological entity recently described, about two decades, and not very much known, having as synonyms the terms Brainstem Processing Syndrome, Central Auditory Dysfunction, Neural Synchrony Disorder[1], and more recently Auditory Dyssynchrony[2], and Auditory Neuropathy Spectrum Disorder[3, 4]. The latter is more appropriate since it is a heterogeneous disorder in both clinical and hearing features, and the associated etiological factors[4]. Its estimated incidence varies widely, it is considered to be 3-11% among children with sensorioneural hearing loss, usually from studies of populations at risk[5].

The incidence in the general population is not well known[6] but is suspected that auditory neuropathy is more common in the infant population than previously thought[7].

It is characterized, in terms of audiology, by the loss of the synchronous activity of the auditory nerve, resulting in altered nerve function associated with the preservation of cochlear function. It is still unknown the exact location in the auditory canal of the retrocochlear lesion[5].

The cases of auditory neuropathy are characterized clinically by: hearing loss, usually bilateral, but unilateral cases was reported[1], in varying degree; functional normality of outer hair cells; evidenced by the presence of Otoacoustic Emissions (OAE) and/or CM[1]; abnormal evoked potentials starting with the wave I of Brainstem Auditory Evoked Potential (BAEP), poor perception of speech and absence of acoustic reflex for ipsilateral and contralateral tones at 110 dB[8].

* Corresponding author:
luzienedb@ig.com.br(Luziene D. B. de Oliveira)
Published online at http://journal.sapub.org/ijbcs

Rance, G. et al, who studied 14 children with AN showed that those patients typically had normal frequency resolution and varying degrees of temporal abnormality which appeared to affect both temporal resolution/amplitude modulation detection and the temporal aspects of frequency discrimination. The severity of this temporal disruption was correlated to speech perception[9].

Thus, adult patients report being able to hear sounds, despite difficulties in the interpretation of speech[10]. The speech recognition in noise is generally poor[11].

However, the Auditory Neuropathy Spectrum Disorder can be diagnosed at any age[1].

The aetiology of auditory neuropathy seems to be multifactorial[10]. Among the proposed causes for auditory neuropathy in children are: genetic disorders, with may include mutations in the otoferlin (OTOF) gene[12], hyperbilirubinemia, neural "immaturity" or other central nervous system disorders[6]. It has also been made associations among auditory neuropathy and infections (measles, mumps), metabolic processes (diabetes, hypoxia), neoplasms (acoustic neuroma) and prematurity[13]. Other probable risk factors are: use of ototoxic drugs and mechanical ventilation in the neonatal period[7] as well as congenital brain abnormalities, perinatal intracranial haemorrhage[4].

There are also cases of patients with genetically determined neurological diseases associated with peripheral neuropathy and hearing loss, for example, Charcot-Marie-Tooth disease[14] or cases: Leber's hereditary optic neuropathy, autosomal dominant optic atrophy, autosomal recessive optic atrophy, Fredreich's Ataxia, Mohr-Tranebjaerg syndrome, Refsum Disease and mitochondrial disease[4]. In adults, merit citation demyelinating conditions such as multiple sclerosis and HIV infection[4].

It is extremely important to obtain the differential diagnosis between classic sensorioneural hearing and auditory neuropathy, especially in children, which require not only a quick and accurate therapeutic intervention with a view to taking advantage of neuroplasticity, known to be higher in this age group, but also the fact of rehabilitation procedure to be different in two cases.

The patients require management to their auditory and communication problems different from those used with patients with classic sensorineural hearing loss[11, 15].

It is worth noting that the newborns without risk factors are generally submitted only to the OAE test, called "ear test or auditory development test", which is not capable of detecting idiopathic cases of auditory neuropathy, since in these cases the OAEs are generally present resulting in delay of the diagnosis[5].

The evolution of this disorder is much variable and probably depending on the underlying cause, some babies improve and start to hear and develop speak until two year old. Other infants stay the same, while some get worse[16].

This disorder can be significant deleterious effect for infants and young children on language development and academic achievement[17].

The successful rehabilitation of speech in such cases depends on early treatment[18].

There isn´t consensus about the effective treatments for children with auditory neuropathy. The hearing health experts' opinions differ about the potential benefits of hearing aids, cochlear implants, and other technologies. May be hearing aids added to personal listening devices such as frequency modulation (FM) systems could be helpful for some children. However, there is no guarantee that an individual with auditory neuropathy might benefit from a hearing aid or cochlear implant. However truth is that a better option is work with a team of professionals who considers the situation and options for each child as well as the child's family and caregivers[16].

For infants with this disorder, however, electrophysiological methods may do not predict auditory detection thresholds and the behavioral response to sound observed by clinicians and parents must to guide the hearing aid fitting decision[17].

2. Objectives

To evaluate the audiological and neurological profile in children with auditory neuropathy, from tertiary health care hearing of the SUS (Public Health Assistence).

3. Methods

We performed retrospective analysis of children's history 0-12 years from Hearing Health Program from December 2005 to December 2010.

3.1. Procedures

Were selected 12 children with suspected AN. We performed a review in all cases searching for results of OAEs, tympanometry, and BAEP, and tonal audio behavioural and/or tonal audiometry.

The neurologic anamnesis were revised aimed possible etiologic factors for AN as neonatal hypoxia, prematurity, sepsis with the use of ototoxic drugs in the neonatal period, and bilirubin encephalopathy.

3.2. Inclusion Criteria

Presence of otoacoustic emissions with tympanometry type A, cochlear microphonic in the Brainstem Auditory Evoked Potential (BAEP) with varying degree loss, and behavioral audiometry or tonal audiometry better than the expected based on the results of BAEP.

3.3. Exclusion Criteria

Were excluded children with completely normal hearing assessment (without sensorioneural deafness) and children with classic sensorioneural hearing loss.

3.4. Analysis of Results

We perform a descriptive statistics of obtained results.

4. Results

The sample comprised 12 children, predominantly male, 83.3% (n = 10), and chronological age 2-12 years (average = 5.4 years).

After tonal audiometry, it was found that sensorioneural hearing loss was ranging from mild to profound (n = 7), as well as the inconstancy of response during the assessment. For children who failed to respond to pure tone audiometry (n=5), an auditory behavioral assessment that was performed, called attention to sound and CER absent. Of these, three laterally located the sound source and one showed an indirect location downward and upward, considered below in chronological age.

It was observed in the impedance, tympanometric curve type A in 60% (n = 7), and others varying between Ar and C in isolated ears, all with ER absent. The TOAE were present in only 4 patients.

In BAEP we found no response with the presence of CM, observed from the change of polarity (condensation and rarefaction) of click stimulus at 99 dB in the registry, 100% cases. Of these, two patients showed CM unilateral, one right and one left.

Regarding to the aetiological factors and clinical history, only four children had no neonatal complications that could explain audiological findings, three other patients had prematurity, hypoxia (3 cases), bilirubin encephalopathy (1 case) or sepsis (5 cases) with the use of ototoxic drugs in the neonatal period. There was a case of chickenpox at 15 months of life and a case associated with transient ataxia at age 3. Only two cases showed no other neurological abnormalities. Four children had epilepsy; three had cerebral palsy, while three others showed hyperkinetic behaviour.

All children were under speech and language interventions.

5. Discussion

The auditory neuropathies vary in several points including age of onset, etiology, presence of peripheral neuropathy, and at physiological and behavioral assessments of auditory function[19].

In the present study of the auditory assessment, variability of responses was observed in the auditory tone audiometry, with inconsistent responses, development of listening skills below the chronological age. It was detected prevalence of both TOAE and BAEP absent, with presence of CM, CER and ER absent.

According to Starr et al[20], AN is characterized by a cochlear integrity with preserved TOAE and CM in all patients with BAEP and ER absent. Rapin and Gravel[13] also describe the verification of the OAE and CM in the diagnosis of auditory neuropathy with absent BAEP. Besides the described features, an auditory discrimination is found disproportionate to the results of tone audiometry[21], with bilateral hearing loss[22].

With the audiological evaluation, it was observed in a tonal audiometry, a response variability characterized by sensorioneural hearing loss ranging from mild to profound (n = 7), and also the inconstancy of response during the assessment. For those children who failed to respond to pure tone audiometry (n = 5), an auditory behavioural evaluation was tested, showing attention to sound and CER absent. Of these, three laterally located sound source, and one of them presented indirect location downward and upward, considered below in chronological age. This way, characterizing an inconsistency of responses to pure tones with variability in degrees of hearing loss (mild to profound) and behavioural responses less than expected for chronological age.

The results of evaluations showed how heterogeneous the clinical demonstrations can be, also the possible aetiological hypothesis in the AN cases. The variability of the chronological age from 2 to 12 years in this study, reinforces the importance of the precocious diagnosis, by giving importance to family`s and caretaker`s complaints and always based on a number of adequate auditory evaluations in order to start a more precocious intervention.

This is in accordance with Dunkley, et. al[6] and Mohnd Khairi, and Zaharah Normastura[22] in their studies that reinforce the importance of the association between TOAE and BAEP in order to allow the diagnosis of auditory neuropathy. Then, without an adequate examination battery it would not be possible to conclude the diagnosis of auditory neuropathy.

Colm Maden et al.[8] analysed the clinical and audiological features of auditory neuropathy in 22 patients with average age of 17 months (9 male and 13 female). Out of these, 20 had the OAE present and BAEP absent or only with CM on BAEP; 2 patients were identified with CM on BAEP and OAE failure. From these data one might think that with increasing age the OAE becomes absent like what was found in our study. But it differs from the findings in our study with 12 children when we observed TOAEs present only in 4 patients. It is consistent with the findings of BAEP with no responses and with the presence of CM, noticed in all cases studied. Of these, two patients showed unilateral CM, one on the right and one on the left. Thus, we observed a higher incidence of absence of TOAE and CM presence on BAEP in children with auditory neuropathy. The absence of TOAE can be justified by the chronological age 2 and 12 years (average = 5.4 years) of children studied.

Thus, the CM research was characterized as an instrument of great trust to carry out the differential diagnosis of AN /

AD. It was concluded that high levels of hyperbilirubinemia at birth seems to be an important risk factor for the occurrence of AN / AD, since the auditory system proved to be susceptible to the neurotoxic effects of this substance. The data in this study regarding the lack of responses on BAEP, CM presence and absence of acoustic reflex validate our findings.

Colm Madden, et. al[8] who studied 22 children point out that factors such as prematurity, genetics and hiperbilerrubinemia appear significantly in AN.

It is worth noting that in our study, concerning the aetiological factors and clinical history, only four children had no neonatal complications, other 3 patients had prematurity, hypoxia (3 cases), bilirubin encephalopathy (1 case) or sepsis (5 cases) with the use of ototoxic drugs in the neonatal period. There was a case of chickenpox at 15 months of life and a case associated with transient ataxia at age 3. Only two cases showed no other neurological abnormalities. Four children had epilepsy; three had cerebral palsy, while three others showed hyperkinetic behaviour.

6. Conclusions

Audiological assessment in children with Auditory Neuropathy Spectrum Disorder shows that the findings are characterized by variability of response on tonal audiometry, with inconsistent responses, development of listening skills below the chronological age. Prevalence of absent TOAE and BAEP absent, with presence of CM, absent CER and ER and incompatibility of responses among the tests when compared to psychoacoustic and auditory behaviour responses are relevant to be mentioned. In addition, importance should be given to parents or caregivers' complaints. It is worth emphasizing that variability of auditory responses in children is mostly informed and questioned by the ones responsible for them. Thus, we conclude that the tests should never be analysed separately.

The findings agree with those in literature, suggesting that auditory neuropathy represents a heterogeneous disorder, being of congenital cause or acquired. Among the causal factors, stand out sepsis with the use of ototoxic drugs, hypoxia and prematurity which sometimes coincide in the same patient. Despite the improvement in neonatal care, hyperbilirubinemia still figures as a cause of hearing impairment and cerebral palsy in our country.

Finally, we emphasize the need to disclose the existence of this nosological entity for health and education professionals, to enable early diagnosis and intervention in those cases, this way, avoiding that those patients are misunderstood / discriminated because they seem to simulate an actual hearing loss, preventing prejudice against those children and their families.

ACKNOWLEDGEMENTS

We thank Terezinha Rosetti for her great help with the English version of this work.

This work was not financially supported by any drug company or funding agency.

REFERENCES

[1] Anastasio, A. R. T.; Freitas Alvarenga, K. F.; Costa-Filho, O. A. Extratympanic electrocochleography in the diagnosis of auditory neuropathy/auditory dyssynchrony. Braz. J. Otorhinolaryngol 74 (1), pp 132-136, 2008.

[2] Hood, L. J.; Berlin, C. I.; Boderlon, J.; Rose, K. Patients with auditory neuropathy/dys-synchrony lack efferent suppression of transient evoked otoacoustic emissions. J Am Acad Audiol, 14 (6), pp. 302-313, 2003.

[3] Saluja, S.; Agarwal, A.; Kler, N.; Amin, S. Auditory neuropathy spectrum disorder in late preterm and term infants with severe jaundice. Int. J. Pedriatr. Otorhinolaryngol, 74, pp.1292-1297, 2010.

[4] Manchaiah. V. K. C.; Zhao, F.; Danesh, A. A.; Duprey, R. The genetic basis of auditory neuropathy spectrum disorder. Int. J. Pedriatr. Otorhinolaryngol, 75, pp.151-158, 2011.

[5] Domingues, F. J. R.; Herrero, J. D. C.; Gallardo, N. C.; Aguilera R, P. Prevalence of auditory neuropathy: prospective study in a tertiary-care center. Acta Otorrinolaringol Esp, 58(6), pp. 239-245, 2007.

[6] Dunkley, C.; Farnsworth, A.; Mason, S.; Dodd, M.; Gibbin, K. Screening and follow up assessment in three cases of auditory neuropathy. Arch Dis Child, 88, pp. 25-26, 2003.

[7] Rance, G.; Beer, D. E.; Cone-Wesson, B.; Shepherd, R. K.; Dowell, R. C.; King, A. M.; Rickards, F. W.; Clark, G. M. Clinical Findings for a Group of Infants and Young Children with Auditory Neuropathy. Ear & Hearing, 20 (3), p238, 1999.

[8] Madden C.; Rutter, M.; Hilbert, L.; Greinwald, J. H.; Choo, D. I. Clinical and audiogical features in auditory neuropathy. Arch Otolaryngol Head Neck Surg, 128, pp. 1026-1030, 2002.

[9] Rance, G.; Mckay, C.; Grayden. Perceptual Characterization of Children with Auditory Neuropathy. Ear & Hearing, 25 (1), pp 34-46, 2004.

[10] Zeng, F. G.; Oba, S.; Garde, S.; Sininger, Y.; Starr A. Temporal and speech processing deficits in auditory neuropathy. Neuroreport, 10, pp. 3429-3435, 1999.

[11] Hood, L. J. Auditory neuropathy: What is it and what can we do about it? The hearing journal, 51 (8), pp. 10-18, 1998.

[12] Varga, R.; Kelley, P. M.; Keats, B. J.; Starr, A.; Leal, S. M.; Cohn, E.; Kimberling, W. J. Non-syndromic recessive auditory neuropathy is the resultof mutations in the otoferlin (OTOF) gene. J Med Genet, 40, pp. 45–50, 2003.

[13] Buchman, C. A.; Roush, P. A.; Teagle, H. F. B.; Brown, C. J.; Zdanski, C. J.; Grose, J. H. Auditory neuropathy characteristics in children with cochlear nerve deficiency. Ear & Hearing. 27, pp. 399-408, 2006.

[14] Rapin, I.; Gravel, J. "Auditory neuropathy": physiologic and pathologic evidence calls for more diagnostic specificity. Int. J. Pedriatr. Otorhinolaryngol, 67, pp. 707-728, 2003.

[15] Doyle, K. J.; Sininger, Y.; Starr, A. Auditory neuropathy in childhood. The Laryngoscope, 108(9), pp. 1374-1377, 1998.

[16] Online Available: http://www.nidcd.nih.gov/health/hearing/pages/neuropathy.aspx

[17] Online Available: http://www.childrenscolorado.org/conditions/speech/danielscenter/ANSD-Guidelines.aspx

[18] Tapia, M. C.; Lirola, A.; Moro, M.; Candela F, A. Auditory neuropathy in childhood. Acta Otorrinolaringol Esp, 51(6), pp. 482-489, 2000.

[19] Starr, A.; Sininger, Y. S.; Pratt, H. The varieties of auditory neuropathy. J. Basic. Clinic Physiol Pharmacol, 11(3), pp. 215-230, 2000.

[20] Starr, A.; Picton, T. W.; Sininger, Y. S.; Hood, L.J.; Berlin, C. I. Auditory neuropathy. Brain, 119, pp. 741-753, 1996.

[21] Vlastarakos, P.V.; Nikolopoulos, T. P.; Tavoulari, E.; Papacharalambous, G.; Korres. Internacional Journal of Pediatric Otorhinolaryngology, 72, pp. 1135-1150, 2008.

[22] Khiari, M.; Normastura, A.; Zaharah, W. Auditory neuropathy: three cases among a group with sensorioneural hearing loss. Singapore Med J, 50 (9), pp. 324, 2009.