Pure-Tone Hearing Thresholds and Brainstem Auditory Evoked Potentials in Sporadic Ataxia

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Int Arch Otorhinolaryngol 2020;24(1):e86–e92.

Abstract

Introduction Spinocerebellar ataxia (SCA) is part of a genetic and clinical heterogeneous group of neurodegenerative diseases characterized by progressive cerebellar ataxia.

Objective To describe the results of audiological and electrophysiological hearing evaluations in patients with sporadic ataxia (SA).

Methods A retrospective cross-sectional study was carried out with 11 patients submitted to the following procedures: anamnesis, otorhinolaryngological evaluation, tonal and vocal audiometry, acoustic immittance and brainstem auditory evoked potential (BAEP) tests.

Results The patients presented with a prevalence of gait imbalance, of dysarthria, and of dysphagia; in the audiometric and BAEPs, four patients presented with alterations; in the acoustic immittance test, five patients presented with alterations, predominantly bilateral.

Conclusion The most evident alterations in the audiological evaluation were the prevalence of the descending audiometric configuration between the frequencies of 2 and 4 kHz and the absence of the acoustic reflex between the frequencies of 3 and 4 kHz bilaterally. In the electrophysiological evaluation, the patients presented changes with a prevalence of increased I, III and V wave latencies and the interval in the interpeak I-III, I-V and III-V. In the present study, it was observed that auditory complaints did not have a significant prevalence in this type of ataxia, which does not occur in some types of autosomal recessive and dominant ataxia.

Keywords
- spinocerebellar degenerations
- hearing
- hearing disorders
- audiometry
- evoked response audiometry

received
January 3, 2019
accepted
June 6, 2019

DOI https://doi.org/10.1055/s-0039-1693676.
ISSN 1809-9777.

Introduction

Ataxias are a group of neurodegenerative diseases featuring the presence of progressive cerebellar disorder, followed by several neurological signs and symptoms, such as balance and motor coordination disorders, besides the presence of eye dysfunctions.[1,2]

Ataxias can be divided in six large groups: a) autosomal recessive hereditary ataxias; b) autosomal dominant hereditary ataxias; c) congenital ataxias; d) X-associated ataxia syndrome; e) mitochondrial ataxias; and f) sporadic ataxias (SAs).[3–5] The last type is the group of interest in the present study.

Sporadic ataxia is classified as a rare neurological condition, of late onset, usually in individuals $>$ 40 years old, with no family history, and with a wide range of potential causes. Among them, cerebellar degeneration, alcohol abuse, paraneoplastic syndrome, heavy metal poisoning (toxic reaction), dysfunction of the neuroimmune system, E, B1, B12 vitamin deficiency, infectious diseases (neurosyphilis, Whipple disease, Lyme disease, and human immunodeficiency virus [HIV]), and degenerative diseases can be pointed out.[4,5]

Drumond et al[6] pointed out the complexity of the differential diagnosis of SA, as well as the difficulty in determining its etiology due to its heterogeneity. When its onset occurs after the age of 50 years old, a wide investigation cannot be enough to detect its etiology; therefore, it can be classified as late-onset idiopathic cerebellar ataxia.

Sporadic adult-onset ataxia (SAOA) of unknown etiology is a nonhereditary disorder, distinct from multiple system atrophy. Despite its unknown cause, it is considered as a set of disorders comprising a common clinical syndrome. Epidemiological studies have found prevalence rates ranging from 2.2 to 8.4 per 100,000 individuals. About a third of the SAOA patients have polyneuropathy or affection of the pyramidal tract observed with cerebellar ataxia, although cognitive impairment is not common or present in a mild degree. Neuropathological and imaging studies have shown isolated cerebellar cortical degeneration and mild brainstem atrophy, although there is no established therapeutics for SAOA.[4]

The neurobiological hearing components involve a complexity of events, as well as broad interrelationships in the central nervous system (CNS).[7]

Assessment of the peripheral and central hearing systems is performed by means of behavioral, electroacoustic, and electrophysiological tests. Peripheral hearing involves the amplification and conduction of sound waves, as well as the perception of sound vibrations that are changed into nervous impulses. Central hearing involves the conduction of nervous impulses by means of the ear pathway to the hearing cortex, where they will be coded and recoded, thus gaining linguistic meaning. Brainstem auditory responses will be assessed by means of the electrical activities in the auditory nerve. The effects caused by degenerative processes may involve the central auditory nervous system (CANS).[8]

In some types of cerebellar ataxia, Ikeda et al, Mahmoud, and Zeigelboim et al[9–11] had a large percentage of subjects with hearing loss verified through the audiological evaluation. Some studies,[11–13] using electrophysiological tests in patients with cerebellar ataxia, already verified abnormalities in brainstem auditory evoked potentials (BAEPs) in $>$ 50% of the evaluated subjects, while other studies[14,15] evidenced abnormality in 71 and 46.5%, respectively, in the BAEP assessment of autosomal recessive and dominant spinocerebellar ataxias. In the acoustic immittance measurement, Zeigelboim et al[11] reported disorders in 46.6% of the sample of patients with neurodegenerative diseases, and 44.1% of abnormality in patients with autosomal dominant spinocerebellar ataxia,[15] mainly related to acoustic reflexes.

Due to the audiological findings found in the literature on the alterations observed in ataxia patients, the present study aimed to describe the results of audiological and electrophysiological screening in patients with SA, which is classified as a rare neurological condition.

Methods

The research protocol was approved by the Ethics Committee on Research Involving Human Subjects (registration number: 832.502/2014) at Platform Brazil. All of the examinations were performed after informed consent forms were obtained from all of the participants.

A retrospective cross-sectional study was carried out, assessing 11 patients (4 females and 7 males) diagnosed with SA referred by the Department of Internal Medicine of the Clinical Hospital for assessment in the Department of Otoneurology of a teaching institution in the same city, diagnosed with SCA, sporadic form. Their diagnosis was performed by means of their clinical history, neurological and imaging screening, and genetic testing using polymerase chain reaction (PCR).[16–18]

The age of the patients ranged from 35 to 58 years old (46.6 $\pm$ 8 years old). The time span of the disease ranged from 4 to 13 years (8.8 $\pm$ 2.8 years), as shown in ►**Table 1**.

The evaluated patients did not report a history of exposure to noise nor the use of ototoxic drugs. They reported a family history of presbycusis (two cases), of hypertension (six cases), and of diabetes (three cases).

Patients with auditory disorders and difficulty in understanding simple commands, which would hinder the screening, were excluded from the research.

All of the patients underwent an analysis of their clinical history and an otorhinolaryngological screening.

Audiological evaluation – The patients were submitted to pure tone air conduction threshold audiometry, frequencies from 0.25 to 8 kHz; pure tone bone conduction threshold audiometry, frequencies from 0.5 to 4 kHz; speech recognition threshold (SRT) and speech discrimination score (SDS). For these tests, the Madsen Itera II equipment (Otometrics A/S, Taastrup, Denmark), TDH-39 heaphones (Telephonics Corp., Farmingdale, NY, USA), and thresholds in dBH were used. The equipment was calibrated according to ISO 8253. The level and type of hearing loss were analyzed according to Davis et al and Silman et al.[19,20]

Brainstem auditory evoked potential – This test used 2 channels with a *click* stimulus in 90 dBHL, alternate polarity with a presentation frequency of 21.1 c/s, window of 15 ms,

Table 1 Sporadic ataxia aspects

Case	Age (years old)	Gender	SARA	Disease duration (years)
1	58	M	22.5	12
2	36	F	16	7
3	43	M	10	12
4	37	M	8.5	8
5	46	F	11.5	10
6	49	M	13	13
7	54	F	7	8
8	35	F	14	6
9	48	M	12.5	10
10	54	M	20.5	4
11	53	M	10	7

Abbreviations: F, female; M, male; SARA, scale for the assessment and rating of ataxia.

30 to 3 kHz filter, and at least 2,000 stimuli, and 2 rounds of reproduction. Kendall Medi-Trace 2000 electrodes were placed on the right and left mastoids, and Fz (10–20 system) and ground electrodes were used on the forehead. Clicks were presented by 3A insert earphones. Wave latencies I, III and V, and interpeak intervals I-III, III-V, I-V were analyzed according to Hall.[21] The equipment used was Bio-logic Evoked Potential NavPRO-ONE System (Otometrics A/S, Taastrup, Denmark).

Acoustic immittance evaluation – This procedure has been performed to access the integrity of the middle ear according to Jerger.[22] The acoustic immittance equipment used was Madsen Otoflex 100 (Otometrics A/S, Taastrup, Denmark).

For the statistical analysis, the Fisher exact test was applied, aiming to compare the results of the audiological testing, of the BAEP and acoustic immittance measurement (analyzing normal and abnormal results), and to relate the audiological findings, BAEP and accoustic immitance measurement to the following variables: duration of the disease, age and Scale for the Assessment and Rating of Ataxia (SARA scale). The null hypothesis rejection rule was established at 0.05 or 5%. The Statistica 13.1 software (Dell Software Inc., Round Rock, TX, USA) was used.

Results

The most reported complaints in the anamnesis were: gait imbalance (81.8%), dysarthria (63.8%). and dysphagia (54.5%). Dizziness, dysphonia, tremor, fall, and irradiated pain to the shoulders and the arms were reported by 45.4% of the patients. Tinnitus and hearing loss were referred by 36.3% of the patients, as shown in ►**Table 2**.

The application of the Fisher exact test evidenced a statistically significant difference in the proportion of cases with gait imbalance when compared with tinnitus ($p = 0.0402$) and with hearing loss ($p = 0.0402$), which were the most evidenced symptoms.

Table 2 Frequency of symptoms of 11 patients with sporadic ataxia

Symptoms	Number of patients	Frequency (%)
Gait imbalance	9	81.8
Dysarthria	7	63.8
Dysphagia	6	54.5
Dizziness	5	45.4
Dysphonia	5	45.4
Tremor	5	45.4
Fall	5	45.4
Pain, irradiated to shoulder, arm	5	45.4
Extremities tingling	4	36.3
Tinnitus	4	36.3
Hearing loss	4	36.3
Anxiety	3	27.2
Blurred vision	3	27.2
Diplopia	3	27.2
Fatigue	3	27.2
Depression	3	27.2
Insomnia	2	18.1
Taste alteration	1	9.0

In the audiological evaluation, 4 patients (36.4%) had altered results, with bilateral prevalence, as shown in ►**Table 3**.

The Fisher exact test was used to compare the frequency of alterations in the auditory thresholds of subjects with SA. No statistically significant difference was found between the frequency of ears that presented alterations in the auditory thresholds and of those that presented normal auditory thresholds ($p = 0.1974$).

The results from the SRT and SDS matched the tonal thresholds.

Regarding the BAEP assessment, 4 patients (36.4%) evidenced altered testing: 18.2% bilaterally, and 18.2% in the left ear, according to ►**Table 3**.

The alterations observed in BAEP were: increase of the latency of wave I (case 11), of wave III (cases 6, 9 and 11), of wave V (cases 1, 6, 9 and 11), and of the interpeak intervals I-III, IV, and III-V (cases 1, 6, 9 and 11).

The application of the Fisher exact test showed a statistically significant difference between the frequency of ears that presented alterations in BAEP and those that presented normal auditory results in this evaluation ($p = 0.0030$).

In the acoustic immittance measurement, 5 patients (45.4%) evidenced abnormal testing, with bilateral prevalence, as shown in ►**Table 4**.

The Fisher exact test was used to compare the frequency of alterations in acoustic immittance of subjects with SA. No statistically significant difference was found between the

Table 3 Results of audiological and brainstem auditory evoked potential evaluations in 11 patients with sporadic ataxia

Patients	Audiological evaluation		Brainstem auditory evoked potential	
	Right ear	Left ear	Right ear	Left ear
1	Down-sloping hearing loss beginning at 2kHz	Down-sloping hearing loss beginning at 2kHz	Abnormal	Abnormal
2	Normal	Normal	Normal	Normal
3	Normal	Normal	Normal	Normal
4	Normal	Normal	Normal	Normal
5	Normal	Normal	Normal	Normal
6	Down-sloping hearing loss beginning at 4 kHz	SNHL moderate	Normal	Abnormal
7	Down-sloping hearing loss beginning at 4kHz	Down-sloping hearing loss beginning at 4kHz	Normal	Normal
8	Normal	Normal	Normal	Normal
9	Normal	Normal	Abnormal	Abnormal
10	Normal	Normal	Normal	Normal
11	Down-sloping hearing loss beginning at 4kHz	SNHL severe	Normal	Abnormal

Abbreviation: SNHL, sensorineural hearing loss.

Table 4 Results of the acoustic immittance test in 11 patients with sporadic ataxia

Patients	Acoustic immittance evaluation			
	Right ear		Left ear	
	Tympanometric	Acoustic reflex	Tympanometric	Acoustic reflex
1	Type A	Absent in the 3 and 4kHz frequencies	Type A	Absent in the 3 and 4kHz frequencies
2	Type A	Present	Type A	Present
3	Type A	Present	Type A	Present
4	Type A	Present	Type A	Present
5	Type A	Present	Type A	Present
6	Type A	Absent in the 4kHz frequency	Type A	Absent
7	Type A	Absent in the 4kHz frequency	Type A	Absent in the 4kHz frequency
8	Type A	Present	Type A	Present
9	Type A	Absent in the 3 and 4kHz frequencies	Type A	Absent in the 3 and 4kHz frequencies
10	Type A	Present	Type A	Present
11	Type A	Absent in the 3 and 4kHz frequencies	Type A	Absent

frequency of ears that presented alterations in this test and those that presented normal auditory results ($p = 0.5000$).

The relation between the audiological findings, BAEP, and acoustic immittance with variables for age, disease duration, and SARA scale can be observed in ►**Table 5**.

The application of the Fisher exact test verified no statistically significant difference between the audiological finding, BAEP and acoustic immittance with variables for disease duration ($p = 0.6515$), ($p = 0.1970$) and ($p = 0.3918$) age ($p = 0.0879$), ($p = 0.4697$) and ($p = 0.1970$) and SARA scale ($p = 0.6182$), ($p = 0.3818$) and ($p = 0.7273$).

Discussion

The anamnesis showed the prevalence of gait imbalance (81.8%) and of dysarthria (63.8%) as the most reported complaints by the patients. As for tinnitus and hearing loss (36.3%), they were less reported by the patients, showing that they were not the main complaint among this population. These findings corroborate the study by Bürk et al,[12] who reported gait imbalance and dysarthria as the most relevant complaints, observed in 100% of the cases.

In the present research, only 36.4% of the researched patients evidenced hearing impairments. Disregarding no significant difference, a higher proportion of altered cases was observed among older patients, as well as a longer time span of the disease. In relation to the SARA scale, there was a higher proportion of impairments among patients who scored higher, that is, with worse degrees of ataxia. This result matches the reported complaints of the patients, in which auditory issues did not have a significant prevalence.

Ikeda et al[9] assessed several types of ataxia, with atrophy of multiple systems in the cerebellar prevalence, cortical cerebellar atrophy, and hereditary ataxias, including SCA 31,

Table 5 Distributions of audiological, brainstem auditory evoked potential, and acoustic immittance test in 11 patients with sporadic ataxia

Audiology findings, *n*	**Disease duration (years)**		**TOTAL**	***p-value***
	4–8	**9–13**		
Abnormal	2	2	4	0.6515
Normal	4	3	7	
TOTAL	6	5	11	
Audiology findings, *n*	**Age (years old)**		**TOTAL**	***p-value***
	35–49	50–58		
Abnormal	1	3	4	0.0879
Normal	6	1	7	
TOTAL	7	4	11	
Audiology findings, *n*	**SARA (score)**		**TOTAL**	***p-value***
	≤ 9	10–22.5		
Abnormal	1	3	4	0.6182
Normal	1	6	7	
TOTAL	2	9	11	
BAEP findings, *n*	**Disease duration (years)**		**TOTAL**	***p-value***
	4–8	**9–13**		
Abnormal	1	3	4	
Normal	5	2	7	0.1970
TOTAL	6	5	11	
BAEP findings, *n*	**Age (years old)**		**TOTAL**	***p-value***
	35–49	**50–58**		
Abnormal	2	2	4	
Normal	5	2	7	0.4697
TOTAL	7	4	11	
BAEP findings, *n*	**SARA (score)**		**TOTAL**	***p-value***
	≤ 9	**10–22.5**		
Abnormal	–	4	4	
Normal	2	5	5	0.3818
TOTAL	2	9	11	
Acoustic immittance findings, *n*	**Disease duration (years)**		**TOTAL**	***p-value***
	4–8	**9–13**		
Abnormal	2	3	5	
Normal	4	2	6	0.3918
TOTAL	6	5	11	
Acoustic immittance findings, *n*	**Age (years old)**		**TOTAL**	***p-value***
	35–49	**50–58**		
Abnormal	2	3	5	
Normal	5	1	6	0.1970
TOTAL	7	4	11	
Acoustic immittance findings, *n*	**SARA (score)**		**TOTAL**	***p-value***
	≤ 9	**10–22.5**		
Abnormal	1	4	5	
Normal	1	5	6	0.7273
TOTAL	2	9	11	

Abbreviation: BAEP, brainstem auditory evoked potential; SARA, scale for the assessment and rating of ataxia.

and observed that the audiogram results did not show any significant difference or specificity in the impaired hearing levels among the ataxic groups, averaging 25.6 dB in the right ear and 29.2 dB in the left ear in the auditory assessment, corroborating the fact that the hearing issue was not their main complaint. As for Friedreich ataxia, an autosomal recessive inherited disease, studies showed that 90% of the patients had hearing loss, considering this result as an important finding in the differential diagnosis for cerebellar ataxias.[10] For Zeigelboim et al,[11] who assessed 30 patients with Friedreich ataxia, audiometric testing unveiled 43.3% of altered cases, with bilateral prevalence in 36.7% of them.

The evaluation of the neurophysiological integrity of the brainstem by the BAEP is due to the synchronicity of the neural element, which can be observed by the wave overlap, adequate morphology, wave latency, and interpeak intervals. The deterioration of the function of the nervous system, most of the time, occurs initially through the cortex, the subcortical regions, and, finally, the brainstem.[23]

Biacabe et al and Nachmanoff et al[24,25] indicate that, in neurodegenerative diseases, the most evidenced auditory dysfunctions are observed in the BAEP and occur in the inferior colliculi, in the lateral lemniscus, and in the cochlear nucleus.

In the BAEP assessment, in the present study, abnormalities were observed in 36.4%, prevalence of latency increase in waves I, III and V, and in the interpeak intervals I-III, I-V and III-V. Bürk et al[12] assessed 104 patients with SA and observed prominent axonal neuropathy in the electrophysiological testing, with reduced amplitude in 50% of the patients, and abnormal BAEP in 58.3% of the cases. In another study, Sinatra et al[13] assessed 22 patients (15 with familial ataxia and 7 with SA), and evidenced altered BAEP in 72.7% of the cases. The authors did not find any correlations with the variables for age, gender, and time span of the disease, and reported that the result did not change after 15 and 24 months of follow-up. The authors suggested that the involvement of the auditory pathways must have a genetic cause. Zeigelboim et al[11] assessed 30 patients with Friedreich ataxia and evidenced abnormalities in 56.6% of the cases, which were bilateral in 50%.

Knezevic et al and Zeigelboim et al [14,15] evidenced abnormality in 71 and 46.5% of the cases, respectively, in the BAEP assessment of autosomal recessive and dominant spinocerebellar ataxias. These authors found important abnormalities in the brainstem auditory pathways in these types of ataxias.

In the acoustic immittance measurement, we observed 45.4% of disorders. There is scarce literature for that finding. Zeigelboim et al[11] reported disorders in 46.6% of the 30 assessed cases. However, in neurodegenerative diseases, the cochlear nucleus is affected, which may interfere in the mechanism of the acoustic reflex.[26] In autosomal dominant spinocerebellar ataxia, studies reported 44.1% of abnormalities.[15] In the present study, a greater abnormality was observed in the acoustic immittance measurement, specifically in the acoustic reflex investigation.

We emphasize the limited size of the sample as a consequence of the difficulty of attending patients to the place where the study was performed, due to the difficulty of locomotion of these subjects, as well as the need of an accompanying person to assist the patient to attend the service. Another limitation is related to the rare frequency of this type of ataxia in the population, as well as to the high mortality rate of those who have this condition. We suggest studies with a greater number of subjects, as well as longitudinal surveys for an auditory follow-up of those with sporadic ataxia.

Conclusion

The most evidenced abnormalities in the audiological testing were the prevalence of the descending audiometric configuration at frequencies between 2 and 4 kHz, bilaterally, and the absence of the acoustic reflex at the frequencies of 3 and 4 kHz, bilaterally.

In the electrophysiological assessment, abnormalities were observed with prevalence in the latency increase of waves I, III and V, as well as interpeak intervals I-III, I-V and III-V.

Conflicts of Interests
The authors have no conflicts of interests to declare.

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