

Marília Mendes Silva<sup>1</sup> Roberto Paulo Correia de Araújo<sup>1</sup> Fernando Antonio Glasner da Rocha  
Araujo<sup>2</sup> Julia de Souza Valente<sup>1</sup> Ana Paula Corona<sup>3</sup> **Keywords**Scleroderma, Systemic  
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Perda Auditiva**Correspondence address:**Marília Mendes Silva  
Instituto de Ciências da Saúde,  
Universidade Federal da Bahia – UFBA  
Avenida Reitor Miguel Calmon, s/n,  
Vale do Canela, Salvador (BA), Brasil,  
CEP: 40110-100.  
E-mail: silva\_mariliam@hotmail.com

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## Hearing alterations in systemic sclerosis

*Alterações auditivas na esclerose sistêmica***ABSTRACT**

**Purpose:** Describe hearing complaints and alterations in individuals with systemic sclerosis (SS) and to verify the development of audiological manifestations. **Methods:** This is a cross-sectional study with a prospective phase, conducted in the period from 2012 to 2015, with patients with medical diagnosis of SS. Sociodemographic data, year of disease onset, year of diagnosis and disease subtype were collected. Later, audiological anamnesis was performed to identify complaints and symptoms and to investigate the performance of audiometry before the study and, after that, a basic audiological evaluation was conducted. **Results:** Fifty individuals participated in the study. Dizziness and tinnitus were the most frequent symptoms. Hearing loss was identified in 23 (46%) individuals; most of them were of sensorineural type, of variable degrees and configurations. The analysis of hearing thresholds obtained in the audiological evaluation performed in 2012 and, later, in 2015, indicated onset or progression of hearing loss, with aggravation of 10dB in most frequencies evaluated, being more expressive in acute frequencies. **Conclusion:** High rate of hearing complaints and alterations in individuals with SS and onset and/or progression of hearing loss in those who underwent serial audiological evaluation were observed.

**RESUMO**

**Objetivo:** Descrever as queixas e alterações auditivas em indivíduos com esclerose sistêmica (ES), bem como verificar a evolução do quadro audiológico. **Método:** Trata-se de estudo seccional, com uma fase prospectiva, realizado no período de 2012 e 2015, com pacientes com diagnóstico médico de ES. Foram coletados dados sociodemográficos, ano de início da doença, ano de diagnóstico e subtipo da enfermidade. Posteriormente, foram realizadas a anamnese audiológica, para identificação de queixas e sintomas e para a investigação de realização de audiometria progressa ao estudo, e, em seguida, a avaliação audiológica básica. **Resultados:** Participaram do estudo 50 indivíduos. Tontura e zumbido foram os sintomas mais frequentes. A perda auditiva foi identificada em 23 (46%) indivíduos, sendo a maioria do tipo sensorineural, de grau e configurações variáveis. A análise dos limares auditivos obtidos na avaliação audiológica realizada em 2012 e, posteriormente, em 2015 indicou desencadeamento ou progressão da perda auditiva, com piora de 10dB na maioria das frequências avaliadas, sendo mais expressiva nas frequências agudas. **Conclusão:** Elevada frequência de queixas e alterações auditivas em indivíduos com ES e desencadeamento e/ou progressão da perda auditiva naqueles que realizaram avaliação audiológica sequencial.

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<sup>1</sup> Instituto de Ciências da Saúde, Universidade Federal da Bahia – UFBA - Salvador (BA), Brasil.

<sup>2</sup> Faculdade de Medicina, Hospital Universitário Prof. Edgard Santos, Universidade Federal da Bahia – UFBA - Salvador (BA), Brasil.

<sup>3</sup> Departamento de Fonoaudiologia, Universidade Federal da Bahia – UFBA - Salvador (BA), Brasil.

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## INTRODUCTION

Systemic sclerosis (SS) is a complex multisystemic disorder, with abnormalities in the immune system, connective tissue and vascular system. It is characterized by chronic inflammation, diffuse small vessel damage and progressive fibrosis in the skin and in various organs<sup>(1-4)</sup>.

The incidence of SS was estimated at 0.6-122 per million people per year, and prevalence rates range from 4 to 489 cases per million individuals<sup>(5,6)</sup>. SS occurs in several geographic areas, with regional incidence differences, and in different races, with higher incidence rate in blacks compared to whites. Additionally, women are affected three times more than men and the average age of disease onset is between 30 and 50 years<sup>(1,2,5,7,8)</sup>.

The clinical classification of SS has been recognized in two main categories, using the extent of skin involvement as discriminator: diffuse, which is characterized by generalized skin involvement, in addition to early involvement of internal organs; and limited, in which the skin involvement is usually restricted to fingers, forearm and face and late involvement of internal organs<sup>(3)</sup>.

International investigations have described audiovestibular alterations in individuals with SS. In addition, they reveal a variable prevalence regarding the degree and type of hearing loss and, in only one study, the relationship between hearing and disease type and severity was verified<sup>(9-13)</sup>. However, the relationship between hearing loss and SS is not fully understood and none of the investigations previously conducted have presented prospective design. In Brazil, only the previous publication of partial results of the present study was located<sup>(14)</sup>.

In this context, and considering that SS is a progressive disease with symptomatology that affects the social life of individuals, in addition to the biological plausibility of auditory alteration by the pathophysiology of this disease, the aim of this study was to describe auditory complaints and alterations in individuals with SS, and to verify the evolution of the audiological condition.

## METHODS

This is a cross-sectional epidemiological survey, allied to a prospective phase, developed with all patients followed at a referral outpatient rheumatology clinic of a university hospital in the State of Bahia. SS diagnosis was established according to the international criteria of the American College of Rheumatology (ACR) - European League Against Rheumatism (EULAR) of 1980 and 2013 and data collection was carried out in two different moments in the year 2012 and 2015<sup>(15,16)</sup>. The year of the first audiological evaluation was considered the zero time (T0) of the study. Time one (T1) was established as the year in which individuals performed audiological reevaluation.

In both moments, identification data (name, age and date of birth) and information regarding the year of onset of symptoms, year of diagnosis and SS subtype were collected through analysis of medical records. Subsequently, audiological anamnesis was performed to identify lifestyle, auditovestibular complaints and symptoms, as well as sociodemographic data and previous audiometry (audiometry prior to participation in the study).

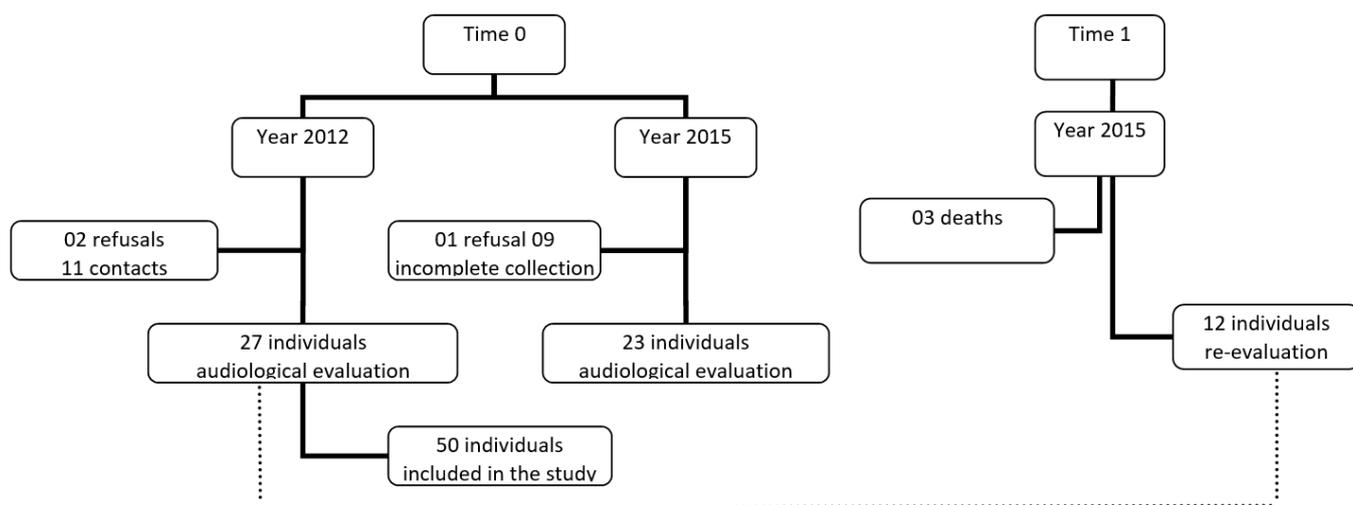
In the audiological anamnesis, subjects were questioned about the presence of complaints related to hearing. The first reported complaint was considered as the main complaint. The following questions were asked regarding the presence of the following symptoms: hearing loss, difficulty in understanding speech, otalgia, otorrhea, discomfort to loud sounds, autophony, tinnitus and dizziness. If the individual reported hearing loss, the characteristics of the condition (unilateral or bilateral), onset and progression were investigated. For those who reported presence of tinnitus or dizziness, type, intensity and frequency of occurrence were characterized.

Regarding health and lifestyle conditions, individuals were questioned about exposure to noise and chemical products, presence of other comorbidities, hospitalization with risk of death, head trauma, chemotherapy and / or pulse therapy and consumption of ototoxic drugs and smoking. For those who reported smoking, it was considered yes for current smokers and ex-smokers, with consumption time greater than or equal to two years, and no for those who never smoked. Subsequently, all subjects underwent basic audiological evaluation and those who presented auditory thresholds above 25dBNA, at any frequency searched, unilaterally or bilaterally, were considered as having hearing loss. The type of hearing loss was classified as sensorineural, conductive or mixed, and the degree was stratified into five categories: mild, moderate, moderately severe, severe and profound. The hearing loss configuration was classified as horizontal, descending, inverted U-curve and notch<sup>(17)</sup>. Hearing losses that did not meet the classification regarding degree and configuration were considered as not classifiable.

Data collected were typed and organized with the help of the EpiData 3.1 software. Initially, mean, median, standard deviation, interquartile range, minimum value and maximum reference value and frequencies of sociodemographic variables were estimated. Otoneurological symptoms and findings were then described and quantified. In addition, in the prospective analysis to identify the audiological evolution of individuals who were evaluated in T0 and T1, a comparison was made between audiometric findings obtained in both times. The presence of change in thresholds was considered when the difference between T1 and T0 was equal to or greater than 10dB at least in one frequency, considering that lower variation among audiometric thresholds may occur due to factors such as ambient noise, temperature, transducers used, motivation of patients and examiner's criteria<sup>(17)</sup>.

Fifty individuals with medical diagnosis of SS participated in the study. In 2012, the Rheumatology Service registered the follow-up of 40 individuals with SS, and anamnesis and audiological evaluation were performed in 27 of them. In 2015, this service followed 84 individuals, of whom only 61 were active patients, that is, they attended medical appointments previously scheduled by the service. Of the 61 active patients, 23 were included in the study. In 2015, audiological reevaluation of 12 of the 27 individuals already included in the study in 2012 was also performed (Figure 1).

This study was approved by the Research Ethics Committee of the Health Sciences Institute, through protocol No. 943.022, and all participants signed the Informed Consent Form.



**Figure 1.** Flowchart of inclusion of individuals with systemic sclerosis in the study

## RESULTS

Of the 50 individuals aged 26-76 years and mean age of 49.25 years (SD = 15.68), 41 were of females. As for self-reported skin color, two participants declared themselves as white, 30 as brown and 18 as black. Table 1 shows the characteristics of the disease, health conditions and lifestyle of individuals with SS. Of the 50 individuals with SS, 35 had the disease for a period of more than four years and most did not present risk factor that could contribute to the onset / progression of hearing loss. In relation to health conditions, it was verified that 25 individuals reported the presence of at least one comorbidity in addition to SS at the time of anamnesis, and arterial hypertension was the most frequent (14). However, nine individuals had no comorbidity or risk factor due to hearing loss.

Regarding the presence of self-reported hearing complaints, it was verified that 27 individuals had at least one complaint, being more frequently the report of hearing loss (16). Regarding previous investigation of auditory acuity, it was observed that 41 of the 50 subjects who participated in the study had never performed basic audiological evaluation. The audiovestibular symptoms of individuals with SS are presented in Table 2. It was observed that dizziness was the most frequently reported symptom, followed by tinnitus. Dizziness was characterized by the majority as discrete and sporadic and in 62.9% of participants, it presented a rotational character. In relation to tinnitus, most individuals characterized it as tonal, with acute pitch and sporadic occurrence.

The findings of the audiological evaluation revealed that 27 of the 50 subjects had normal hearing and 23 were diagnosed with hearing loss of variable degree and configurations (Table 3). The majority of hearing losses was of sensorineural type, with greater impairment in the acute frequencies. Regarding the results of the acoustic immittance measurements, most individuals presented type A tympanometric curve and contralateral stapedial acoustic reflex with non-suggestive differentials of Metz target recruitment (equal to or below 60dB)<sup>(18)</sup>.

**Table 1.** Disease characterization, presence of risk factors for hearing loss and lifestyle of individuals with systemic sclerosis

Variables	N (%)
SS subtype	
Limited	22 (44)
Diffuse	13 (26)
No information	15 (30)
Exposure to noise	8 (16)
Exposure to chemicals	9 (18)
Other comorbidities	25 (50)
Hospitalization with risk of death	3 (6)
Chemotherapy	1 (2)
Pulse Therapy	12 (24)
Cranial Trauma	8 (16)
Ototoxic drugs	11 (22)
Smoking	20 (40)

Caption: N = Number of individuals; SS = Systemic Sclerosis. Source: Research data

**Table 2.** Presence of audiovestibular symptoms of individuals with systemic sclerosis

Variables	N (%)
Hearing loss	20 (40)
Hearing loss	21 (42)
Otalgia	22 (44)
Otorrea	3 (6)
Autophony	20 (40)
Tinnitus	32 (64)
Dizziness	35 (70)
Discomfort to intense sounds	17 (34)

Caption: N = Number of individuals. Source: Research data

When comparing the information of individuals who underwent audiological evaluation at T0 and reevaluation at T1, no changes were observed in sociodemographic characteristics, disease characteristics, health conditions and lifestyle. Regarding audiovestibular symptoms, the number of individuals who reported hearing loss increased from four to six, comparing the two hearing moments evaluated (T0 and T1). In relation

**Table 3.** Characterization of hearing loss and immitanciometric findings in individuals with systemic sclerosis

Variable	RE	LE
	N (%)	N (%)
Hearing Loss	22 (95.7)	21 (91.3)
Type		
Conductive	1 (4.5)	2 (9.5)
Sensorineural	20 (90.9)	18 (85.7)
Mixed	1 (4.5)	1 (4.8)
Degree		
Mild	2 (9.1)	1 (4.8)
Moderate	1 (4.5)	2 (9.5)
Moderately Severe	-	1 (4.8)
Severe	1 (4.5)	-
Not classifiable	18 (81.9)	17 (80.9)
Configuration		
Horizontal	1 (4.5)	3 (14.3)
Descending Light	1 (4.5)	2 (9.5)
Descending accentuated	1 (4.5)	-
Inverted U-Curve	1 (4.5)	-
Notch	2 (9.2)	1 (4.8)
Not classifiable	16 (72.8)	15 (71.4)
Timpanometric Curve		
A Type	21 (95.4)	19 (90.6)
Ar Type	1 (4.6)	1 (4.7)
Ad Type	-	1 (4.7)
Contralateral stapedal acoustic reflex		
Normal	13 (59)	13 (61.9)
Increased	1 (4.5)	1 (4.8)
Decreased	4 (18.2)	4 (19)
Absent	4 (18.2)	3 (14.3)

**Caption:** N = Number of Individuals; RE = Right ear; LE = Left ear

to the other symptoms investigated, no new complaints were recorded and maintenance of the characteristics of symptoms previously reported was observed.

In the analysis of the basic auditory evaluation results, it was verified that immitanciometry findings and type and configuration of hearing loss were maintained. Of the 12 individuals who took part in the study T1, the majority aged up to 55 years (8), five of them self-referred as brown, six as black and one as white. Among the 24 reassessed ears in the year 2015, it was observed that in 16 ears, there was worsening of the auditory threshold by airway and bone, in at least one frequency, and in 3 ears, which had auditory thresholds within normality standards, in all frequencies in the year 2012, some auditory threshold was identified in the audiological reevaluation above 25 dBNA, indicating the onset of hearing loss. In the remaining ears (five), no changes in audiometric thresholds were observed. Considering the hearing loss laterality, it could be observed that in T0 (2012), four right ears and four left ears had hearing loss. In T1, 12 ears were identified with hearing loss, being five right ears and seven left ears (Chart 1 and 2).

Figure 2 shows the distribution of auditory threshold variation per frequency, and ear of the 12 SS individuals who underwent auditory reassessment in T1. The analysis of the variation of thresholds obtained in T0 and T1 revealed a worsening of up to 10dB in most frequencies evaluated, and this worsening was more expressive in acute frequencies.

**Chart 1.** Aerial tonal thresholds of the right ear obtained in the audiological evaluation performed at the different times of the study (T0 and T1)

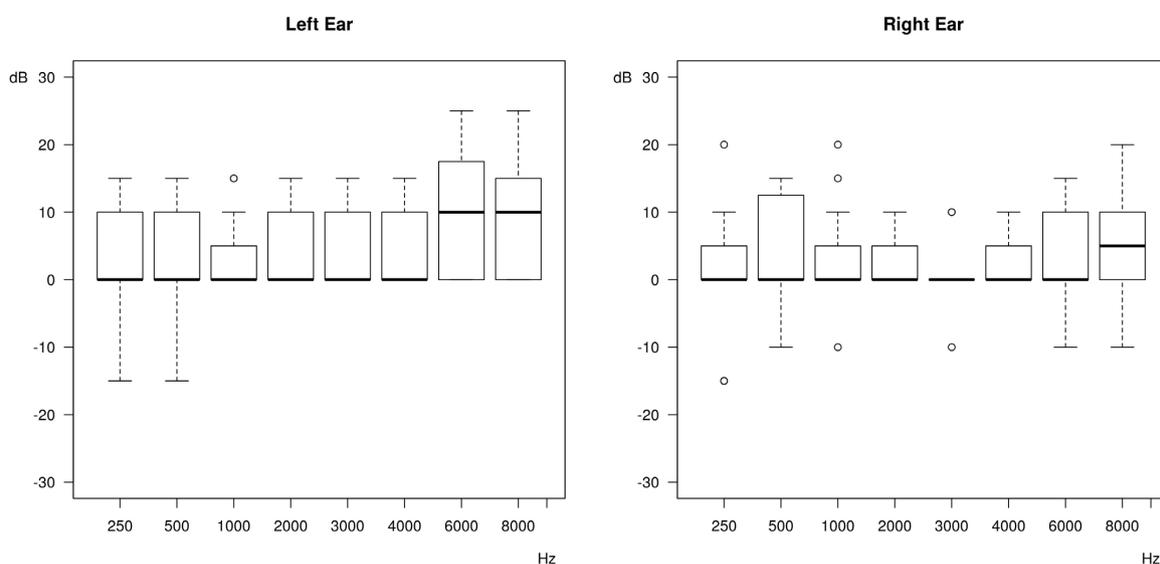
CASE	AGE (years)	TIME IN THE STUDY	FREQUENCIES (Hz)							
			250	500	1000	2000	3000	4000	6000	8000
1	54	T0	25	25	15	20	10	20	30	40
	57	T1	25	25	15	20	15	25	30	40
2	51	T0	10	10	<b>20</b>	<b>5</b>	<b>25</b>	<b>30</b>	45	<b>55</b>
	52	T1	15	10	<b>40</b>	<b>15</b>	<b>35</b>	<b>40</b>	50	<b>65</b>
3	26	T0	25	15	15	5	10	5	5	0
	28	T1	5	20	5	5	10	0	10	0
4	43	T0	<b>15</b>	15	20	20	20	20	35	30
	47	T1	<b>25</b>	20	25	25	10	25	35	30
5	53	T0	10	10	5	10	10	10	15	15
	56	T1	10	15	10	10	10	15	10	10
6	41	T0	<b>10</b>	<b>5</b>	<b>5</b>	10	20	25	<b>20</b>	<b>20</b>
	43	T1	<b>20</b>	<b>20</b>	<b>20</b>	15	25	30	<b>35</b>	<b>40</b>
7	60	T0	<b>0</b>	<b>0</b>	0	<b>15</b>	20	<b>10</b>	10	<b>5</b>
	63	T1	<b>20</b>	<b>15</b>	5	<b>25</b>	25	<b>20</b>	15	<b>15</b>
8	39	T0	20	20	15	15	15	<b>15</b>	<b>15</b>	<b>0</b>
	42	T1	25	25	15	15	15	<b>25</b>	<b>25</b>	<b>15</b>
9	21	T0	25	15	25	15	15	15	20	15
	24	T1	10	20	20	10	15	10	10	15
10	55	T0	20	20	5	5	5	10	10	<b>5</b>
	57	T1	5	10	10	10	5	5	15	<b>15</b>
11	32	T0	30	20	15	15	15	15	<b>15</b>	15
	35	T1	35	25	20	20	20	15	<b>25</b>	20
12	52	T0	25	20	15	<b>10</b>	<b>5</b>	<b>10</b>	<b>10</b>	<b>10</b>
	54	T1	20	25	20	<b>20</b>	<b>20</b>	<b>20</b>	<b>25</b>	<b>20</b>

**Source:** Research data

**Chart 2.** Aerial tonal thresholds of the left ear obtained in the audiological evaluation performed at the different times of the study (T0 and T1)

CASE	AGE (years)	TIME IN THE STUDY	FREQUENCIES (Hz)							
			250	500	1000	2000	3000	4000	6000	8000
1	54	T0	55	<b>45</b>	45	50	45	40	35	50
	57	T1	55	<b>55</b>	45	50	45	<b>55</b>	<b>55</b>	55
2	51	T0	<b>20</b>	<b>15</b>	20	<b>10</b>	<b>30</b>	45	<b>45</b>	<b>50</b>
	52	T1	<b>30</b>	<b>30</b>	25	<b>20</b>	<b>45</b>	45	<b>70</b>	<b>65</b>
3	26	T0	20	10	10	5	5	0	5	10
	28	T1	15	10	15	10	10	0	5	5
4	43	T0	15	15	15	20	15	25	30	35
	47	T1	20	15	15	20	15	20	30	30
5	53	T0	5	10	10	10	5	10	15	10
	56	T1	5	5	5	10	0	10	15	5
6	41	T0	<b>10</b>	<b>5</b>	<b>5</b>	10	15	<b>20</b>	<b>10</b>	<b>0</b>
	43	T1	<b>25</b>	<b>20</b>	<b>20</b>	15	15	<b>30</b>	<b>35</b>	<b>25</b>
7	60	T0	<b>5</b>	<b>0</b>	15	<b>15</b>	20	10	10	<b>0</b>
	63	T1	<b>15</b>	<b>10</b>	10	<b>25</b>	25	15	10	<b>15</b>
8	39	T0	<b>15</b>	15	15	20	<b>15</b>	<b>15</b>	<b>25</b>	15
	42	T1	<b>25</b>	20	15	25	<b>25</b>	<b>25</b>	<b>35</b>	20
9	21	T0	15	15	20	15	5	0	15	<b>5</b>
	24	T1	15	20	20	10	10	10	20	<b>15</b>
10	55	T0	20	20	5	<b>0</b>	0	10	<b>5</b>	<b>0</b>
	57	T1	5	5	10	<b>10</b>	5	5	<b>15</b>	<b>10</b>
11	32	T0	30	20	10	15	10	15	<b>15</b>	10
	35	T1	35	25	15	15	20	20	<b>25</b>	15
12	52	T0	25	20	15	<b>5</b>	10	10	10	15
	54	T1	15	10	15	<b>15</b>	10	15	15	15

Source: Research data



**Figure 2.** Difference in dB of hearing threshold by frequency obtained in the tonal threshold audiometry at Time Zero and Time One (N=12)

## DISCUSSION

The results of this study reveal high frequency of audiovestibular symptoms and hearing loss in individuals with SS. This finding is consistent with results of previous investigations conducted with the purpose of evaluating the audiovestibular impairment in SS patients<sup>(9-14)</sup>.

There is no consensus in literature regarding the complaints most frequently presented by individuals with SS. In the current study, the most common complaint was dizziness, followed by tinnitus. This finding is consistent with studies that also investigated hearing-related complaints in individuals with SS<sup>(12)</sup>. However, other studies have identified hearing loss as the most frequent complaint<sup>(9,10)</sup>.

Dizziness was identified in 35 of the 50 individuals who participated in the present study. It is believed that this symptom is related to the reduction or interruption of blood supply to the vestibular system, through the labyrinth artery, due to vasculopathy, which is one of the clinical manifestations of SS<sup>(10,19)</sup>. The second most frequent complaint was tinnitus, reported by 32 individuals, most of them being tonal and with acute pitch. These tinnitus features are compatible with sensorineural hearing loss and are consistent with the type of auditory alteration identified in individuals with SS<sup>(18,20,21)</sup>.

The frequency of hearing loss in individuals with SS in the present study was 46% and the mean age was 50.56 years. These findings are corroborated by most international studies, which reveal high frequency of auditory alterations in individuals with SS, in the same age group<sup>(9,10,12)</sup>. However, it was verified that the frequency of hearing loss in individuals with SS is higher than expected for individuals without the disease in the same age group<sup>(22)</sup>. In Brazil, epidemiological studies to investigate the frequency of hearing loss by age group are scarce. In a study conducted with the aim of investigating the prevalence of hearing loss in an urban area population, it was identified through tonal audiometry that 84.8% of participants had normal hearing and 15.2% were diagnosed with hearing loss. The authors concluded that the prevalence of estimated hearing loss is in line with international prevalence<sup>(23)</sup>. However, this prevalence was lower than that described in a study conducted with 2,427 people in Rondônia to provide the first population-based data on deafness and hearing loss in Brazil. In this study, hearing loss was verified in 26.1% of the population<sup>(24)</sup>. In this context, it should be considered that only aging does not explain the high frequency of hearing loss in individuals with SS in the current study.

The analysis of the audiological evaluation of individuals with SS in relation to the type of hearing loss revealed high frequency of symmetrical bilateral sensorineural hearing loss with greater impairment of acute frequencies, with variable degree and configurations. These findings are consistent with the type of hearing loss identified in international research conducted with the same purpose<sup>(9,10)</sup>. However, some studies have described less frequently of unilateral mixed or conductive hearing loss<sup>(9,10,12,25)</sup>. Despite the report of this type of impairment, no explanations were found about the biological mechanism of the conductive component involved in hearing loss. It is believed that myopathy, which causes muscle damage, as well as skin alterations and connective tissue fibrosis characteristic of SS, are related to the mechanical alterations of the joints that make up the middle ear ossicle tympanum system and therefore are responsible for modifications in this system<sup>(9,10,14)</sup>.

SS has features that involve connective tissues and vascular systems, causing generalized fibrosis in the skin and vessels, in addition to causing chronic inflammation that may be due to a generalized lesion<sup>(1-3,26,27)</sup>. In this way, the blood tissue is the main target and can cause vasculopathy and, later, collagen deposition may occur. Thus, endothelial dysfunction is implicated in the pathogenesis of this multisystem disease<sup>(9,10,14)</sup>.

Vasculitis can be responsible for the malfunctioning of hair cells in the Corti organ, causing sensorineural hearing loss<sup>(9)</sup>.

In addition, decreased cochlear blood flow may play an important role in hearing loss in individuals with SS due to inflammation of the small vessels of the epineurium or inflammation of the cochlear nerve *vasa vasorum*<sup>(11)</sup>. This event is plausible if there is anatomical alteration of the terminal vessels of the cochlea associated with decreased capillary density, resulting in reduced blood flow and therefore tissue hypoxia<sup>(28)</sup>. Thus, tissue hypoxia causes the death of hair cells in the cochlea, which is an organ highly sensitive to blood changes and, consequently, this change is expressed with sensorineural hearing loss.

The analysis of the health condition and history of exposure revealed that some individuals in the present study presented in the SS course other comorbidities and use of ototoxic drugs that could explain, trigger or intensify hearing loss. The influence of ototoxic drugs on the hearing of individuals with SS has already been reported in other studies. However, the findings of Maciaszczyk et al.<sup>(12)</sup> do not confirm this hypothesis and suggest that the most aggressive course of the disease and immunosuppression therapy does not affect hearing, since when investigating a group of individuals who made use of cyclophosphamide and sodium methotrexate, they found that the use of these drugs had no ototoxic adverse effects. Additionally, the case of a 65-year-old patient diagnosed with bilateral sensorineural hearing loss before the onset of SS was reported. This individual underwent drug therapy (intravenous immunoglobulin - IVIG and increased methotrexate dosage) for SS and, after one month of treatment, symptoms, including hearing loss, improved significantly<sup>(11)</sup>.

The relationship between SS and the development of hearing loss is still obscure, although the disease pathophysiology is consistent with the hypotheses for the biological plausibility of this alteration. Thus, when considering those who reported having no risk factor or comorbidity for hearing loss investigated in the present study, it was verified that five of nine individuals had sensorineural hearing loss. This finding evidences the high frequency of hearing loss in these individuals and corroborates the hypothesis of the possible role of SS in triggering, intensification or worsening of hearing loss.

The analysis of the audiological evaluations of the 12 individuals performed at T0 and T1 revealed triggering or progression of hearing loss. No longitudinal studies that have followed the hearing of individuals with SS were found. However, three international sectional studies, two of which included a comparison group, point out that there is no relationship between the development or worsening of hearing loss and the time of disease in individuals with SS<sup>(9,10,12)</sup>. Another important point to be discussed is the contribution of age in these findings. According to the World Health Organization<sup>(29)</sup>, the age of 60 is considered as a cutoff point for old age. However, although the characteristics of hearing losses identified are similar to alterations due to aging, it was observed that the majority of individuals who underwent auditory reassessment in 2015 aged up to 55 years. In addition, the greatest differences among aerial tonal auditory thresholds identified in T1, compared to T0, were observed in individuals younger than 55 years.

The majority of individuals who participated in the present study (41) did not perform previous hearing assessment.

Considering the high frequency of audiovestibular symptoms and that these can be related to real hearing problems, the diagnostic investigation of these symptoms should be routine in the follow-up services of individuals with SS. This procedure is reinforced by the findings of the investigation conducted to estimate the validity of three unique questions used to assess self-reported hearing loss compared to pure tone audiometry in an adult population. Researchers conclude that each question provides answers with sufficient accuracy to recommend the use of self-reported hearing loss in epidemiological studies with adults<sup>(30)</sup>.

The present study presents limitations, which implied analyses regarding the role of SS in auditory alterations. Among them, the small number of participants who were followed in T1, which did not allow the conduction of logistic regression to determine the contribution of each factor investigated for the triggering or worsening of hearing loss. It is believed that the recruitment of individuals who entered the study at T0, and also in T1, was hampered by the fact that the majority live in the inner state or did not attend the reference service regularly, due to locomotion difficulties caused by the progression of SS. In addition to these factors, the poor prognosis of the disease should be considered, which led to death three patients who participated in the study at T0. Since it is a population with some motor limitations, it is also believed that a greater number of participants would be obtained if audiological evaluation was performed in the same physical space of the medical monitoring service.

However, despite the limitations, the findings of this study contribute to a better understanding of the audiological profile of individuals with SS and contribute with evidence to clarify the role of this disease in hearing loss. In addition, these results may also provide subsidies for health professionals who directly work with individuals with SS to indicate routine audiological assessment. Thus, hearing loss can be early identified and thus provide adequate rehabilitation. In addition, routine audiological evaluation is suggested for individuals with SS since the symptoms of SS are progressive and patients become increasingly debilitated due to other physical restrictions and, consequently, become limited to socialization.

Further studies are necessary for a better understanding of hearing loss in individuals with SS, considering that hearing is a means of integration into social life. Further investigations are suggested with the inclusion of a comparison group, since individuals with the disease in the present study present hearing loss frequency higher than that estimated for individuals without the disease in the same age group. In addition, there are other risk factors that can trigger hearing loss and, therefore, the study with a comparison group may offer subsidies for the investigation of the association between SS and hearing loss. In addition, the investigation of the relationship between clinical and laboratory manifestations of SS and audiological findings should be considered in future studies, since the findings may support the indication of routine auditory evaluation to professionals working in the area, even before the onset of audiovestibular symptoms.

## CONCLUSION

The results obtained in the present study indicate high frequency of otoneurological complaints and auditory alterations in individuals with SS, with reduced report of risk factors for hearing loss, and the onset and / or progression of hearing loss in those who underwent sequential audiological evaluation.

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#### Author contributions

*MMS main researcher, participated in project construction, data collection and article writing; RPCA assisted in article writing; FAGRA assisted in data collection and article writing; JSV assisted in project construction and data collection; APC assisted in project construction, data collection and article writing.*