

# Cochlear implant in Kearns-Sayre syndrome: case study of twin sisters

## Implante coclear na síndrome Kearns-Sayre: estudo de caso de irmãs gêmeas

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### ABSTRACT

Hearing impairment may be related to several factors, including hearing loss due to certain genetic syndromes. Kearns-Sayre syndrome is characterized by mutations in mitochondrial DNA (deoxyribonucleic acid), responsible for energy production (adenosine triphosphate - ATP), which is extremely important for the development of structures that need it, such as the cochlea. The case was followed in the hospital, since 2000, due to the progressive characteristic of the hearing loss observed in the audiological examinations and the findings in cases related to the syndrome. The intervention with individual sound amplification devices (AASI) proved to be of little benefit to the good oral communication of one of the patients, who was diagnosed as having bilateral profound hearing loss. Thus, after discussions at clinical meetings, the team opted for the indication of the cochlear implant for the patient, according to the current criteria for indication of this surgery, and with which it obtained good results. Her twin sister, who presented good results with AASI, will continue to undergo audiological follow-up, to verify the evolution of the case and discuss a new conduct, if necessary. Patients with suspected or diagnosed Kearns-Sayre syndrome should seek the audiological diagnosis, since it is a possible progressive hearing loss, requiring rehabilitation with the use of hearing devices. Maintaining oral communication is extremely important because in these cases other functions will be impaired, such as muscle tone and vision.

**Keywords:** Hearing loss; Cochlear implantation; Syndrome; Diseases in twins; Audiology

### RESUMO

A deficiência auditiva pode estar relacionada com diversos fatores, entre eles, a perda auditiva decorrente de determinadas síndromes genéticas. A síndrome Kearns-Sayre é caracterizada por mutações no DNA (*deoxyribonucleic acid*) mitocondrial, responsável pela produção de energia (trifosfato de adenosina - ATP), que é de extrema importância para o desenvolvimento de estruturas que dela necessitam, como a cóclea. O caso foi acompanhado no hospital, desde o ano 2000, devido à característica progressiva da perda auditiva constatada nos exames audiológicos e nos achados em casos relacionados à síndrome. A intervenção com os aparelhos de amplificação sonora individual (AASI) se mostrou pouco benéfica para a boa comunicação oral de uma das pacientes, que passou a ter o diagnóstico de perda auditiva profunda bilateral. Assim, após discussões em reuniões clínicas, a equipe optou pela indicação do implante coclear para a paciente, segundo os critérios atuais para indicação desta cirurgia, e com o qual obteve bons resultados. Sua irmã gêmea, que apresentou bons resultados com AASI, continuará em acompanhamento audiológico, para se verificar a evolução do caso e discutir uma nova conduta, caso necessário. Pacientes com suspeita, ou diagnóstico de síndrome Kearns-Sayre devem buscar o diagnóstico audiológico, pois se trata de uma possível perda auditiva progressiva, sendo necessária a reabilitação com o uso de dispositivos auditivos. Manter a comunicação oral é de extrema importância, pois, nestes casos, outras funções serão prejudicadas, como o tônus muscular e a visão.

**Palavras-chave:** Perda auditiva; Implante coclear; Síndrome; Doenças em gêmeos; Audiologia

Study performed at the Audiological Research Center of the Hospital for Rehabilitation of Craniofacial Anomalies, Universidade de São Paulo – USP – Bauru (SP), Brazil, as a conclusion work on Multiprofessional Residency in Hearing Health.

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**Conflict of interests:** No

**Authors' contribution:** LSO collects and analyzes data from charts, tabulates data in spreadsheets and, later, graphs, discussion and conclusion of the work; KCB guiding and co-author of the study, assistance in the withdrawal of medical records for data collection, analysis of results and finalization and correction of work; EBO co-author of the study, initial investigation of the case, assistance in the dissemination of medical record data and in the correction of work; KFA co-orienting the project, idealization of the proposal, assistance in the development of the work and corrections for the scientific article.

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**Received:** 1 /16 /2017; **Accepted:** 6 /5 /2017

## INTRODUCTION

Several causes of hearing loss are reported in the literature and, regardless of etiology, it is important that diagnosis and intervention be performed early. Approximately 30% of genetic hearing loss occurs associated with a syndrome<sup>(1,2)</sup>.

Kearns-Sayre syndrome (KS) is related to genetic mutation in mitochondrial DNA (mtDNA). Mitochondria are an intracellular organelle, which has its own genome (DNA)<sup>(2)</sup>, and it is possible that multiple deletions of this genome occur<sup>(3)</sup>. The inheritance of the mitochondrial genome is maternal because, during the course of fertilization, the spermatozoon's tail, which contains the mitochondria, is displaced during penetration into the ovum. In the KS syndrome, part of the zygote mRNA is not formed, however, these mutations can also occur spontaneously<sup>(4)</sup>.

The main function of mitochondria is to provide energy to cells in the form of ATP (adenosine triphosphate) and some organs require more energy and are more affected by cases of mutations in mtDNA. Among these organs are nerve, muscle, optic, endocrine and auditory cells. The cochlea is an organ that requires a lot of energy, so mutations in the cDNA of hair cells can cause sensorineural hearing loss, bilateral, symmetrical and progressive<sup>(2,4,5)</sup>.

The KS syndrome was first described by Kearns and Sayre in 1959<sup>(6)</sup>, in a case report, which presented with external ophthalmoplegia, pigmentary retinopathy and cardiac conduction disorder (CCD). It is a rare syndrome and it is estimated that the onset of cases occurs in 1.6 for every 100,000 individuals<sup>(7)</sup>.

The diagnosis is made through the observation of a triad: progressive external ophthalmoplegia, pigmentary retinopathy and CCD. The first signs and characteristics of the syndrome usually appear before 20 years of age, as observed in the literature<sup>(3,6,7,8,9,10)</sup>. In addition, it is possible to perform the genetic test, mainly seeking information related to DNAm, where multiple deletions or mutations are found, in cases of KS syndrome<sup>(11,12)</sup>.

In the literature<sup>(9)</sup>, there are reports of KS syndrome in twin siblings: there were no risk factors present, and at age 19, both began to present symptoms of the syndrome, such as palpebral ptosis and sensorineural hearing loss.

Regarding progressive hearing loss, the literature recommended the follow-up of patients with this suspicion, at least every two months, to check for progression. The authors also described that electrophysiological exams in children help in the best conduct for the adaptation of individual sound amplification devices<sup>(13,14)</sup>. It is believed that such monitoring is necessary in order to avoid loss of sensory and, consequently, cognitive auditory information, which can lead to delayed auditory and language development.

Few studies related to KS syndrome have focused on the diagnosis of hearing loss and do not correlate this finding in

twin children. However, some authors argue that the most adequate hearing intervention in cases of progressive hearing loss in patients with KS syndrome and others related to mRNA mutations would be the cochlear implant (CI)<sup>(15)</sup>.

The present study aimed to report the case of twin sisters with KS syndrome, from their audiological diagnosis to the intervention.

## Conducting research

The study was initiated after the approval of the Research Ethics Committee of the Hospital of Rehabilitation of Craniofacial Anomalies of the *Universidade de São Paulo*, Campus Bauru: 090311 /2015, CAE 42447215.8.0000.5441. This is a longitudinal descriptive study of twin sisters with KS syndrome and audiological diagnosis of sensorineural hearing loss performed by the interdisciplinary team of a hospital. Cases are described as twin 1 (T1) and twin 2 (T2).

The patients were enrolled in the hospital in June 2000 and have been in follow-up ever since. Therefore, in order to demonstrate more accurately the progression of hearing loss and the results of the intervention, we chose to describe two moments of the evaluation: time of hospital admission: first audiological diagnosis, performed in June 2000, when they were 11 years old; The last consultations reported in the medical record, up to the time of the beginning of the survey, conducted in August 2014 and March 2015 for the T1, and in August 2014 for the T2, both of them, then 25 years of age.

## Patients

The mother of the patients, when registering them at the hospital in June 2000, signed the Informed Consent Term, authorizing the consultation of the medical record data, for scientific purposes. In the year of this study, in 2015, the patients and the family were informed that the research would be carried out by analyzing documents attached to the medical records of the hospital and, thus, confirmed the existing permission for the research development.

## CLINICAL CASE PRESENTATION

### Medical evaluation

The first consultation at the hospital was performed by an otorhinolaryngologist, who, on clinical examination, found complaints and characteristics typical of the KS syndrome. During the anamnesis, the mother reported other evaluations and diagnoses made by physicians in her home city, such as astigmatism, hyperopia, visual fatigue, reduced muscular mass with preserved strength, myopathic face, slight restriction of ocular motricity and alteration in muscle tone.

The patients (T1 and T2) were referred to perform the genetic test to confirm the suspicion of the syndrome, already verified by another institution and informed by the mother, through a document, to the multidisciplinary team, who reported all these data in the medical record, in Form of anamnesis and evolution of the case.

### Audiological diagnosis

#### Twin 1

The data presented below were reported by the mother in a speech-language pathology during the first care in June 2000, when the patient was 11 years old.

- There is no family history of hearing loss and syndromes.
- There were no interurrences during gestation.
- Report of mumps and chickenpox, about 3 years old.
- Neuropsychomotor and language development according to age, however, without previous specialized assessments.
- Hearing complaints began at the age of ten. The T1 patient responded to her name when the voice was very loud and also reported bilateral tinnitus.

Objective tests, such as imitanciometry, transient and distortion product otoacoustic emissions (EOA and DP) and brainstem auditory evoked potential (BERA), and subjective exams such as pure tone audiometry (PTA) were analyzed, and speech perception tests (SPT) (Chart 1).

The results of the subjective exams were compatible with the electrophysiological ones, in both moments of evaluation, as shown in Figure 1.

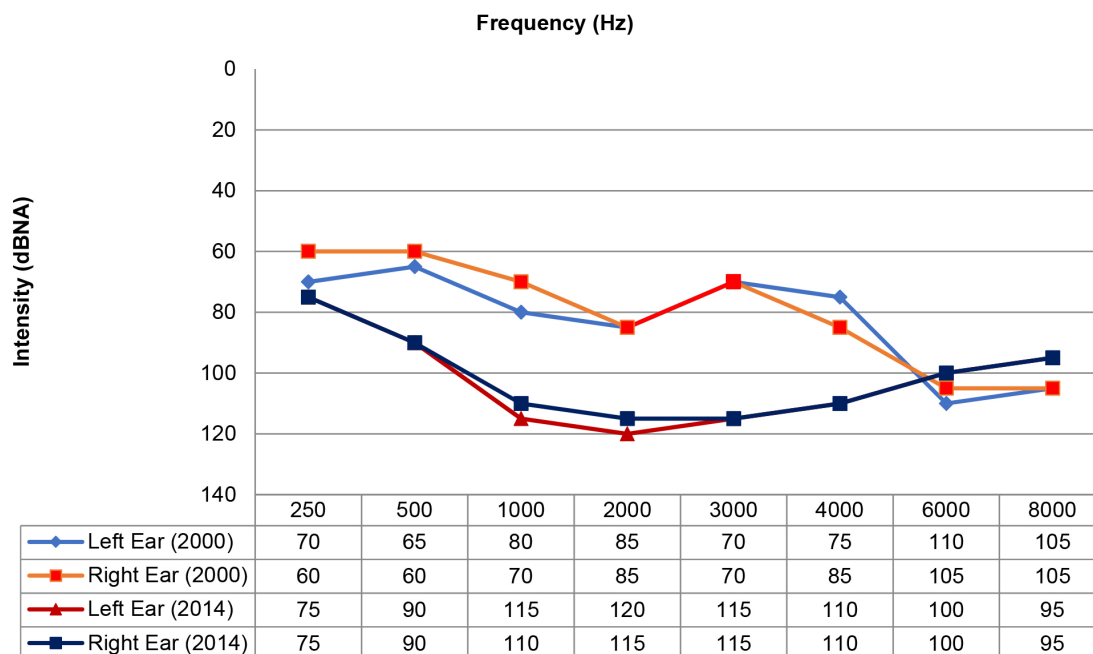
In 2000, after the examinations, the professionals could diagnose hearing loss as bilateral sensorineural hearing loss and thus, adapt the individual sound amplification device and perform the respective validation and verification tests. However, in 2014, there was a significant progression of the degree of hearing loss, which became profound bilateral, requiring replacement of the hearing aid with another, higher and more recent technology, as well as redoing the verification tests (Figure 2).

In the year 2000, the application of the speech perception test (SPT) with individual sound amplification device in all patients was not yet common. They were performed only in those whose cases would be presented at a meeting, to

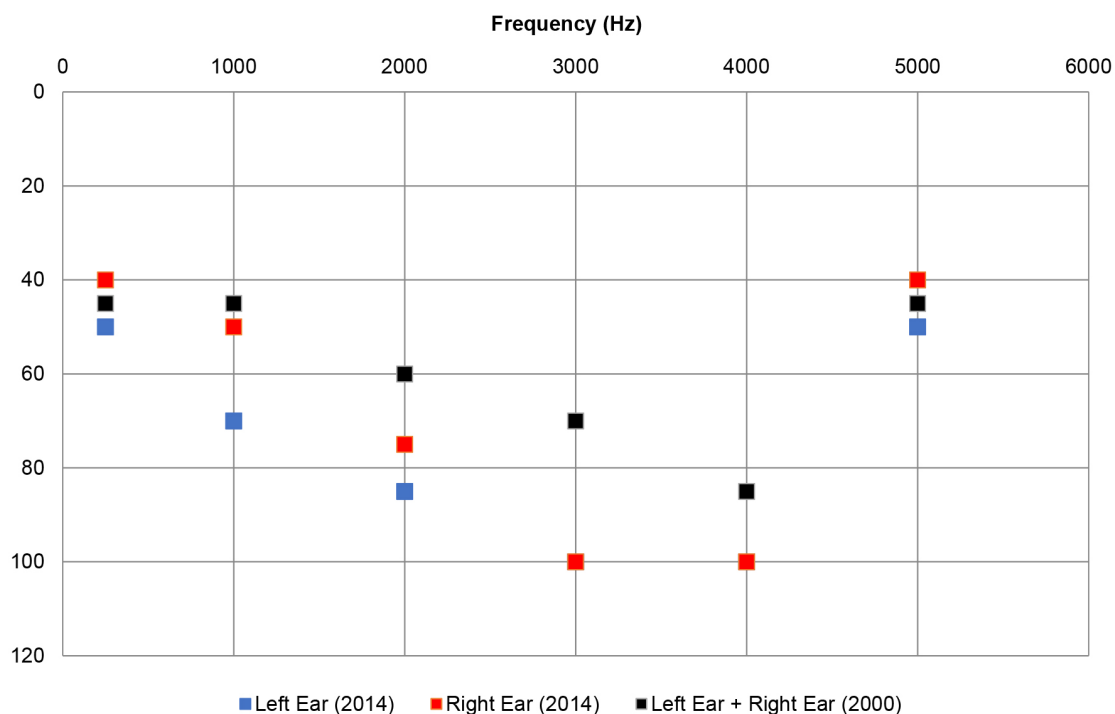
**Chart 1.** Results of the objective examinations of twin 1 (2000 and 2014)

Exam performed	Right ear (RE)	Left ear (LE)
Immittanciometry	Curve type A Reflexes absent in 2000 and 2014	Curve type A Reflexes absent in 2000 and 2014
EOAt e EOAPD	No record in medical records in 2000 Absent in 2014	No record in medical records in 2000 Absent in 2014
BERA click	No record in medical records in 2000 Absent in 2014	No record in medical records in 2000 Absent in 2014

**Subtitle:** EOAt = Transient Evoked Otoacoustic Emissions; EOA = Distortion product evoked otoacoustic emissions; BERA = Auditory Evoked Brain Stem Potential



**Figure 1.** Pure tone audiometry - twin 1



**Figure 2.** Amplified threshold search with Individual Sound Amplification Apparatus - twin 1

discuss cochlear implant surgery (CI). For this reason, in the T1 patient's chart, a SPT evaluation was only found in 2014. The right ear (RE) score was: detection of Ling test sounds for /a/, /u/, /i/ and /m/ = 100% and for /s/ and /ʃ/ = 0%; Name discrimination = 100%; Discrimination of the question affirmation = 0%; Vocabulary extension = 44%; Identification of sentence length = 60% and sentence identification = 0%

In the left ear (LE), the scores found were: detection of Ling test sounds for /a/, /i/, /u/ and /m/ = 100% and for /s/ and /ʃ/ = 0%; Name discrimination = 90%; Discrimination of the question /affirmation = 0%; Vocabulary extension = 28%; Identification of sentence length = 30% and sentence identification = 0%.

Despite the progression of hearing loss, twin 1 had good vocabulary and oral language, since the hearing loss was post-lingual. There were no specific examinations of language in the medical record. It is believed that the reason is that the patient is already in adolescence in 2000 and later in adulthood in 2014, and there are no specific language protocols in the hospital sector to evaluate these age groups.

After the examinations, the case was selected to be discussed by the multidisciplinary team regarding the need for surgery for the cochlear implant (CI). Several criteria were approached, based on the analysis of other professionals, such as social worker, psychologist, otorhinolaryngologist and speech therapist, and based on SUS and hospital criteria at the time.

Regarding the speech-language analysis, the main criterion for indication of surgery was the low result in the tests of speech perception in closed set and the difficulty in open set.

In addition, practitioners were concerned that they would not have other hearing aids options in the future if hearing loss were to progress. Still, the team doctors emphasized that the patient was losing other functions, including vision. Therefore, it was necessary that she maintained a good oral communication, to preserve her quality of life, despite the other difficulties caused by the syndrome.

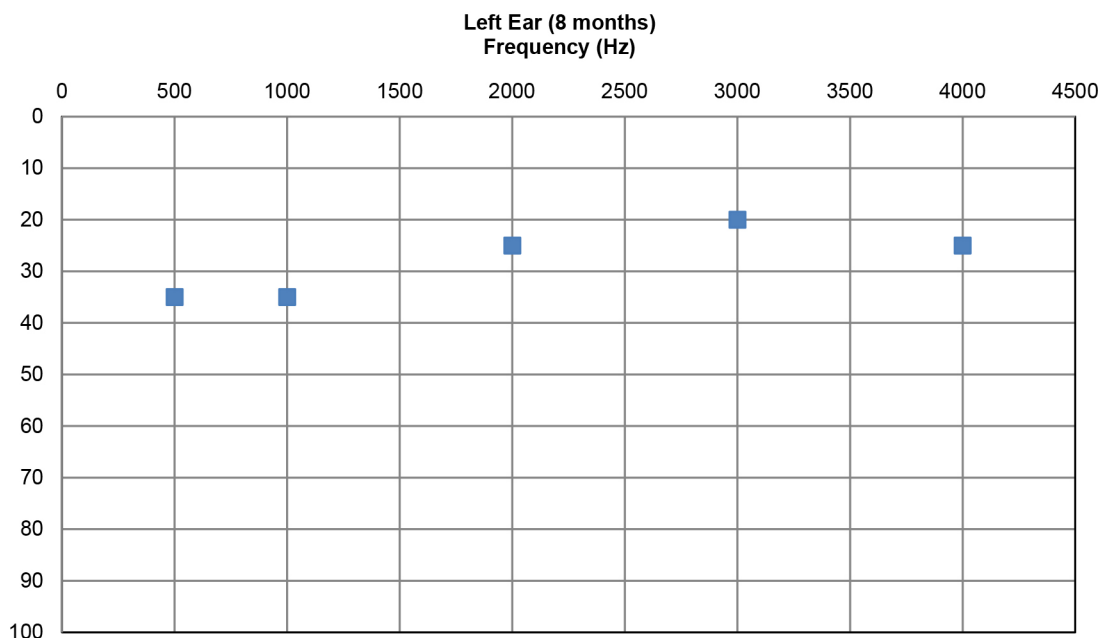
Therefore, we opted for the left side CI surgery, as decided by surgeons. Activation occurred in February 2015 (Figure 3).

Regarding the SPT with CI, performed one month after the activation in March 2015, the patient detected all the sounds of the Ling test, but could not perform other speech tests, complaining that she had not yet become accustomed to all Sounds, because the CI was very different from the individual sound amplification device, which it used for 15 years. The best results appeared eight months after the activation, when the patient performed the SPT in open set, with a result of 45% for the list of sentences. However, regular follow-ups are still being done for new mappings and guidelines.

#### *Twin 2*

The data presented below, as well as for the twin 1, were reported by the mother in a speech-language pathology during the first care in June 2000, when the patient was 11 years old.

- There is no family history of hearing loss and syndromes.
- There were no complications during pregnancy and delivery.
- Contracted mumps and chickenpox near 3 years of age.
- Neuropsychomotor and language development, according to age, however, without previous specialized assessments,
- Hearing complaints began at the age of ten. The T2 patient



**Figure 3.** Amplified threshold search with cochlear implant - twin 1

responded to her name only when the voice was very strong and also reported bilateral tinnitus

Objective tests, such as imitanciometry, transient and distortion product otoacoustic emissions (EOA and DP) and brainstem auditory evoked potential (BERA), and subjective exams such as pure tone audiometry (PTA) were analyzed, and speech perception tests (SPT) (Chart 2).

The results of the subjective exams were compatible with the electrophysiological ones, in both moments of evaluation, as shown in Figure 4.

After all evaluations, in 2000, when twin 2 was 11 years old, the professionals diagnosed the patient with mild sensorineural hearing loss in the left ear and moderate degree in the right ear. Therefore, individual sound amplification device adaptation, compatible with this type of hearing loss, was performed as well as validation and verification tests.

In 2014, when the patient was 25 years old, hearing loss progression was observed, as shown in Figure 4. Thus, the diagnosis changed to bilateral severe grade hearing loss, which made it necessary to Tests to verify their benefits (Figure 5).

In relation to SPT with individual sound amplification

device, performed in 2014, the patient was able to perform all the tests in closed set and obtained a result of 80% in RE and 97% in LE, for the open set tests (list of sentences).

As for patient T1, despite the progression of hearing loss, the T2 patient had good vocabulary and oral language, since hearing loss is post-lingual. There were no specific examinations of language in the medical record. It is believed that the reason is that the patient is already in adolescence in 2000 and later in adulthood in 2014, and there are no specific language protocols in the hospital sector to evaluate these age groups.

The case was also selected for a multidisciplinary meeting, to discuss the CI, including, to compare the results with those of the twin 1. All professionals, such as social worker, psychologist, otorhinolaryngologist and speech therapist, reported the results of their evaluations and, of this information and of the criteria established by SUS and the hospital in 2014, twin 2 were not indicated for CI surgery.

In relation to speech-language evaluation, the main factor for not indicating the CI was a good percentage of correct answers in the SPT with individual sound amplification

**Chart 2.** Results of the objective examinations of the twin 2 hearing (2000 and 2014)

Exam performed	Right ear (RE)	Left ear (LE)
Immittanciometry	Curve type A Reflexes absent in 2000 and 2014	Curve type A Reflexes absent in 2000 and 2014
EOAt e EOAPD	No record in medical records in 2000 Absent in 2014	No record in medical records in 2000 Absent in 2014
BERA click	Absent in 2000 Absent in 2014	Absent in 2000 Absent in 2014

**Subtitle:** EOAt = Transient Evoked Otoacoustic Emissions; EOA = Distortion product evoked otoacoustic emissions; BERA = Auditory Evoked Brain Stem Potential

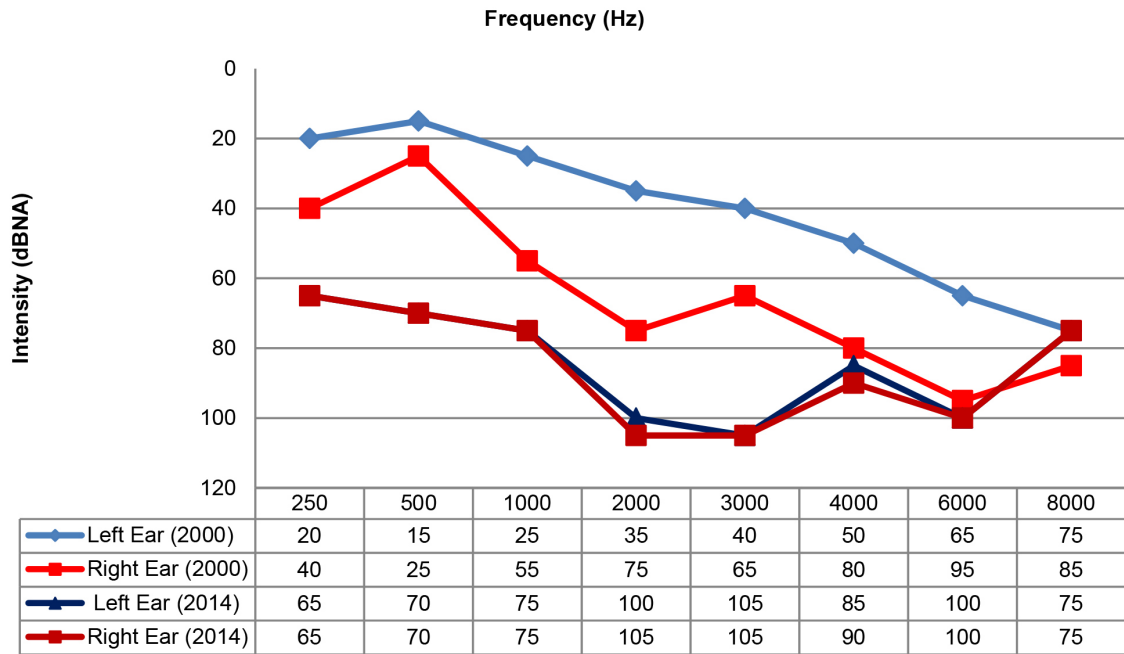


Figure 4. Pure Tone Audiometry - twin 2

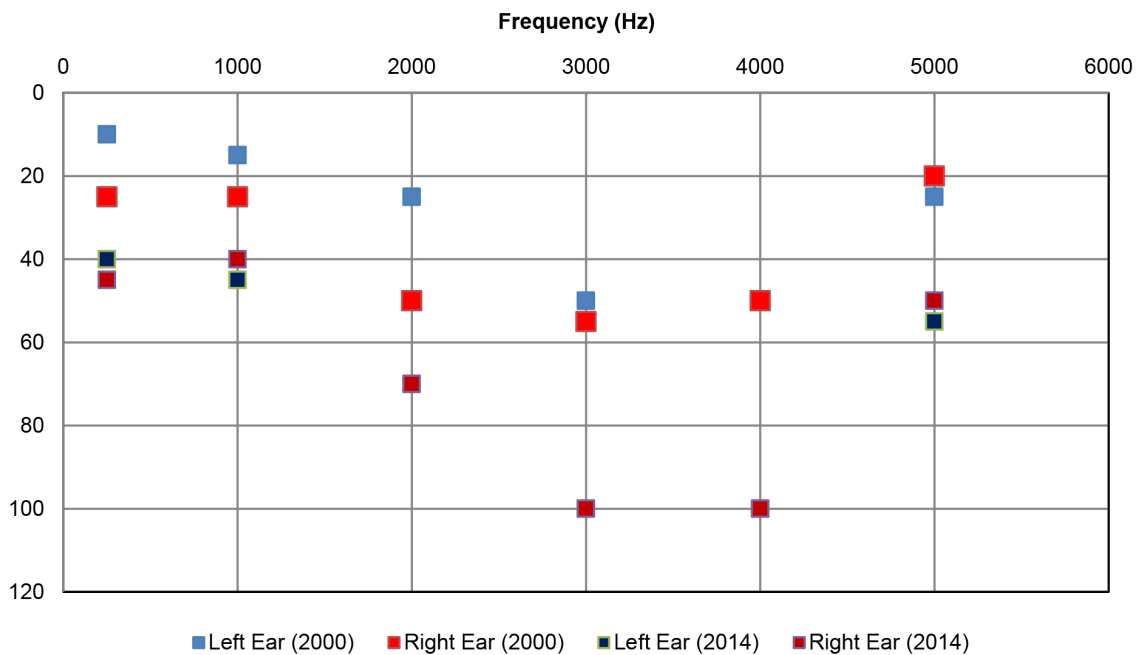


Figure 5. Amplified threshold search with Individual Sound Amplification Apparatus - twin 2

device, demonstrating that the device still provided a benefit. Therefore, it was decided to keep the twin 2 in audiological follow-up, to verify if the hearing loss will progress. Even though this may happen, practitioners believe that there are still other individual sound amplification device replacement options to maintain good oral communication of the patient, which makes it unnecessary at the moment to undergo surgery, since the benefits of CI are very similar to those that individual sound amplification device still provides. However, once that is no longer provides the same benefits, the case of twin 2 will again be discussed by a multidisciplinary team, regarding the

need for CI, to avoid that the progressive loss aggravates the patient's current good communication.

All data referring to the discussion of cases T1 and T2 by the multidisciplinary team were collected in the medical records, from an interview with the professionals responsible for the case.

## DISCUSSION

In the literature<sup>(7)</sup>, the case of twin siblings with KS syndrome was described, demonstrating that there were no



risk factors related to gestation and early childhood, both of which developed normally until the age of 11, when the first complaints of visual acuity altered and hearing difficulties. These findings are compatible with those found in the present study, since the sisters did not present complaints or symptoms until 10 years of age. Other studies<sup>(2,4,5)</sup> also cited the onset of symptoms and diagnosis before the age of 20 years.

In relation to the audiological examinations, the diagnosis of hearing loss was obtained, as well as progression of the auditory thresholds, one of the classic characteristics of hearing impaired patients due to KS syndrome, being, most of the time, sensorineural, symmetrical and progressive<sup>(1,2)</sup>. For this reason, it is believed to be extremely important the audiological diagnosis and early intervention, as well as the monitoring every two or three months, to verify the progression of hearing loss and to define the best behavior in relation to hearing loss adaptation<sup>(13,14)</sup>. Thus, delay in hearing and language development can be avoided.

The etiology of the syndrome is still discussed in the literature. In the case studied (T1 and T2), the only complications during early childhood were diagnoses of mumps and varicella, but these viral infections, when treated, do not usually cause severe sequelae. Therefore, it is believed that the syndrome may have occurred due to genetic mutations.

Another factor that indicated the presence of a genetic mutation in the case of this study was bilateral hearing loss, symmetrical and progressive, as it demonstrates alteration in the cochlea, in agreement with the literature, regarding the genetic mutation of DNAm, which impairs the supply (ATP), especially to the organs that most need it, such as the cochlea<sup>(1,2)</sup>.

In cases of hearing loss caused by mitochondrial diseases, the best intervention option, according to authors<sup>(15,16)</sup>, is the CI, due to the progressive characteristic of these losses. This was also described in this study, in the case of T1, which, even with the individual sound amplification device replacement, after diagnosis of deep hearing loss, did not obtain a hearing benefit verified in the exams, main reason for the indication of CI. In the case of T2, despite the progression of hearing loss, even with the use of hearing aids, from 2000 to 2014, the patient presented favorable speech perception conditions, ie, hearing benefit still existed only with hearing aids, but due to the characteristics Case, the patient will continue to be followed up.

Authors<sup>(15)</sup> reported the case of patients with KS syndrome, who underwent HF. The responses regarding speech recognition and electrophysiological exams with CI were very similar to the responses of CI patients who did not have the syndrome.

It is believed that patients who already use CI (twin 1), together with the contralateral AASI, in addition to speech and language rehabilitation, may present even more satisfactory results in relation to speech recognition, and maintain a good oral communication.

## FINAL COMMENTS

Subjects with suspected KS syndrome should be evaluated for confirmation of the syndrome and early audiological evaluation, because rehabilitation using a hearing aid and / or cochlear implant, together with speech therapy, may bring beneficial results.

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