

## HOW THE SPEECH-LANGUAGE PATHOLOGIST INTERACTS WITH CYSTIC FIBROSIS PATIENTS? A SCOPING REVIEW

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### ABSTRACT

**Objective:** The review describes the changes related to sleep-breathing disorders, communication and feeding processes in subjects with CF, through a scoping review. **Methods:** A scoping review based on the PRISMA protocol was carried out in Pubmed, Lilacs and Scopus databases, to investigate the possibility of including SLP in the treatment of patients with CF. The inclusion criteria included studies that addressed communication and eating processes and disorders related to CF. Studies with a sample that presented other comorbidities that would justify the worsening of the condition, or secondary studies, were excluded. There was no time or language delimitation. **Results:** The search found 1566 works, 27 Lilacs, 1009 Pubmed and 530 Scopus, by analysis of titles, abstract and reading in full. A total of 30 articles were selected for inclusion, 2 Lilacs, 22 Pubmed and 6 Scopus, all related to Speech-Language Pathology. **Conclusion:** It was observed that there is a poor sleep quality due to nocturnal desaturation, mild and moderate

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obstructive apnea. In studies related to hearing, individuals had sensorineural hearing loss. Speech-Language Pathology is of paramount importance in monitoring these subjects.

**KEYWORDS:** Speech-Language Pathology; Cystic Fibrosis; Audiology; Sleep.

### **QUAL A RELAÇÃO DA FONOAUDIOLOGIA COM A FIBROSE CÍSTICA? UMA REVISÃO DE ESCOPO**

#### **RESUMO**

**Objetivo:** Descrever alterações relacionadas aos processos de distúrbios respiratórios do sono, comunicação e alimentação em sujeitos com FC, por meio de uma scoping review. **Métodos:** Foi realizada uma scoping review nas bases de dados Pubmed, Lilacs e Scopus, para investigar a possibilidade de incluir o fonoaudiólogo no tratamento de pacientes com FC, utilizando o checklist PRISMA. Os critérios de inclusão incluíram estudos que abordassem processos de comunicação e alimentação e distúrbios relacionados à FC. Foram excluídos estudos com amostra que apresentasse outras comorbidades que justificassem o agravamento do quadro, ou estudos secundários. Não houve delimitação de tempo ou idioma. **Resultados:** A busca encontrou 1566 trabalhos, sendo 27 Lilacs, 1009 Pubmed e 530 Scopus, por meio de análise de títulos, resumo e leitura na íntegra. Foram selecionados para inclusão 30 artigos, sendo 2 Lilacs, 22 Pubmed e 6 Scopus, todos relacionados à Fonoaudiologia. **Conclusão:** Observou-se que há má qualidade do sono devido à dessaturação noturna, apneia obstrutiva leve e moderada. Em estudos relacionados à audição, os indivíduos apresentaram perda auditiva neurosensorial. A Fonoaudiologia é de suma importância no acompanhamento desses sujeitos.

**PALAVRAS-CHAVE:** Fonoaudiologia; Fibrose Cística; Audiology; Sono.

### **¿CUÁL ES LA RELACIÓN ENTRE LA LOGOPEDIA Y LA FIBROSIS QUÍSTICA? UNA REVISIÓN DEL ALCANCE**

#### **RESUMEN**

**Objetivo:** Describir las alteraciones relacionadas con los procesos de trastornos respiratorios del sueño, comunicación y alimentación en sujetos con FQ, a través de una revisión de alcance. **Métodos:** Se realizó una revisión de alcance en las bases de datos Pubmed, Lilacs y Scopus, para investigar la posibilidad de incluir al logopeda en el tratamiento de pacientes con FQ, utilizando la lista de verificación PRISMA. Los criterios de inclusión incluyeron estudios que abordaran los procesos de comunicación y alimentación y los trastornos relacionados con la FQ. Se excluyeron los estudios con una muestra que presentaba otras comorbidades que justificasen el empeoramiento del cuadro, o estudios secundarios. No había delimitación de tiempo ni de idioma. **Resultados:** La búsqueda encontró 1566 trabajos, de los cuales 27 fueron Lilacs, 1009 Pubmed y 530 Scopus, mediante análisis de títulos, resúmenes y lectura completa. Se seleccionaron 30 artículos para inclusión, 2 Lilacs, 22 Pubmed y 6 Scopus, todos relacionados con Logopedia. **Conclusión:** Se observó que existe mala calidad del sueño por desaturación nocturna, apnea obstructiva leve y moderada. En estudios relacionados con la audición, los individuos tenían pérdida auditiva neurosensorial. La logopedia es de suma importancia en el seguimiento de estos sujetos.

**PALAVRAS-CHAVE:** Fonoaudiología; Fibrosis Quística; Audiología; Sueño.

## 1. INTRODUCTION

Cystic Fibrosis (CF) is a genetic autosomal recessive progressive disorder of the Transmembrane Conductance Regulator (CFTR) gene due to the deficiency, dysfunction or absence of its protein (Morales-Múnera, Rosero-Ascuntar *et al.*, 2020). There are numerous genetic alterations related to CF, but the most common significantly reduces the excretion of chlorine in the human body, so there is an increase in intracellular electronegativity. To try to control this, the body sends a greater flow of Na (Sodium) and water to the cell. This process causes fibrosis resulting from the dehydration of mucous secretions and obstruction of the ducts (Ribeiro, Ribeiro *et al.*, 2002).

In Brazil, the incidence rate of the disease is 1:7 in 576 live births (Athanzio, Silva *et al.*, 2017). Symptoms depend on the affected area, lung disease occurs in more than 90% of the cases, with respiratory symptoms being the most common, such as chronic cough, pneumonia and recurrent bronchitis (Spicuzza, Sciuto *et al.*, 2012).

The treatment of pulmonary disease in CF includes anti-inflammatory drugs, inhaled corticosteroids, antibiotics, mucolytics, hypertonic saline solution and physical therapy (Furlan, Ribeiro *et al.*, 2017). Given the variety of alterations that CF can cause, multidisciplinary treatment is of paramount importance, aiming at a better quality of life and better performance in the physical and psychological aspects (Athanzio, Silva *et al.*, 2017).

Regarding the interface with Speech-Language Pathology (SLP), it can be highlighted that CF basic condition brings interesting correlations, although still little explored. Firstly, the relation with the hearing area is highlighted, considering that the middle ear is directly linked to the respiratory tract through the Eustachian tube, as the buildup of secretions and respiratory infections can cause acute suppurative otitis media (Forman-Franco, Abramson *et al.*, 1979). Recurrent sinusitis and tonsillitis can also occur, presenting anatomical and functional factors that justify the condition of deviated septum, respiratory difficulty, tubal dysfunction, tonsil hypertrophy, adenoids and allergies (Balbani, Sanchez *et al.*, 1998).

Considering these respiratory changes, these patients can acquire mouth-breathing pattern, which can also be related to dental malocclusion and changes in masticatory function (Blacharsh, 1977). In addition to SLP in the adequacy of mastication, SLP focused on breathing has shown significant results, readjusting and raising awareness about respiratory training (Marson, Tessitore *et al.*, 2012).

Still on breathing, Sleep Therapy is an area of expertise recognized by the Brazilian Sleep Association. Therefore, in addition to changes in daytime breathing, the possibility of changing conditions during sleep is highlighted, such as reduced sleep quality due to factors such as: intermittent hypoxemia and transient nocturnal hypercapnia, increased nocturnal respiratory rate and obstructive apnea of sleep (Shakkottai, O'brien *et al.*, 2018).

Breathing is the fuel for vocal production, which can be related to CF due to reduced vocal quality, vocal loudness regardless of gender, and HNR, using acoustic analysis as a parameter to represent the signal-to-noise ratio, increases in the variation of vocal amplitude and frequencies, such as changes in jitter and shimmer, which are normally related to irregularity of vocal fold vibration and, generally, are also related to vocal harshness and the presence of noise in the vocal signal. Turbulence such as partial upper airway obstruction can cause mucus buildup and chronic cough, which are standard symptoms of respiratory pathology in CF patients (Lourenço, Costa *et al.*, 2014).

As exposed above, the CF patient suffers affections in several areas covered by the SLP interaction. A better understanding of what is known, and which are the best therapy approaches for hearing, breathing, mastication and speech in a CF patient will improve their quality of life, reminding that the new medication protocols improved a lot their life expectancy. Thus, a scoping review was carried out to systematically map all the alterations in the cited studies. SLP is an important area in the multidisciplinary work developed for patients with CF, enabling and rehabilitating functions related to communication and nutrition, providing a better quality of life.

The objective of this study was to describe what is known regarding the alterations related to the breathing, communication and feeding processes in subjects with CF, investigating the possibility of SLP therapy, through a scoping review.

## 2. METHODS

This study is a scoping review to investigate the possibility of including SLP in the treatment of patients with CF, by searching the literature in the following databases: Pubmed, Lilacs and Scopus. The scoping review was performed using the PRISMA checklist (Tricco, Lillie *et al.*, 2018), with the studies included in the review, and meeting the inclusion criteria and the review behavior. In this study there were no funding sources.

## 2.1 Inclusion and Exclusion Criteria

The inclusion criteria included studies that addressed communication and eating processes and disorders related to CF. Studies with a sample that presented other comorbidities that would justify the worsening of the condition, or secondary studies, were excluded. There was no time or language delimitation.

## 2.2 Data Collection Procedure

After the searches, the selection was made by reading the titles, and those that were not related to SLP were excluded. Duplicate titles were also excluded and kept only in one of the databases, in total there were 4 duplicates. The others were selected for abstract reading. Research that has not been published in article format, that were not related to SLP, did not have relevant data and literature review were excluded.

Data collection was carried out through a table containing 7 columns, with the topics: "Author, year, country", "Type of study", "Sample", "Instruments applied to measure Speech-Language alterations", "Instrument applied to diagnose CF", "Main results" and "Conclusion".

## 3. RESULTS

After the inclusion and exclusion criteria, 30 articles were selected, showing a variability of published articles on CF, but few studies related to speech-language disorders. The Decs/Mesh terms used in each base and the initial location in the bases are detailed in Table 1.

Table 1 – Results of articles located and selected by database, using search strategies.

Base	Search strategies	Found	Selected by title	Selected by abstract for full reading	Selected for research
Pubmed	("Cystic Fibrosis"[MeSH Terms] OR Mucoviscidosis OR "Pulmonary Cystic Fibrosis") AND ("Stomatognathic System"[MeSH Terms] OR "Mastication"[MeSH Terms] OR "Sleep"[MeSH Terms] OR "Sleep Disorders"[MeSH Terms] OR "Deglutition"[MeSH Terms] OR "Swallowing" OR "Deglutition Disorders"[MeSH Terms] OR "Swallowing Disorder" OR "Dysphagia" OR "Oropharyngeal Dysphagia" OR "Hearing Loss"[MeSH Terms] OR	1009	77	31	22

	"Hypoacusis" OR "Hypoacuses" OR "Hearing Impairment" OR "Voice Quality"[MeSH Terms] OR "Voice"[MeSH Terms] OR "Sleep Apnea, Obstructive"[MeSH Terms] OR "Mouth Breathing"[MeSH Terms] OR "Speech-Language Pathology"[MeSH Terms])				
Scopus	("Cystic Fibrosis" OR Mucoviscidosis OR "Pulmonary Cystic Fibrosis") AND ("Stomatognathic System" OR "Stomatognathic Systems" OR "Masticatory System" OR "Masticatory Systems" OR "Stomatognathic System Abnormalities" OR "Mastication" OR "Chewing" OR "Sleep" OR "Sleeping Habits" OR "Sleep Habits" OR "Sleep Habit" OR "Sleeping Habit" OR "Deglutition" OR "Deglutitions" OR "Swallowing" OR "Deglutition Disorder" OR "Swallowing Disorders" OR "Swallowing Disorder" OR "Dysphagia" OR "Oropharyngeal Dysphagia" OR "Hearing Loss" OR "Hypoacusis" OR "Hearing Impairment" OR "Transitory Deafness" OR "Transitory Hearing Loss" OR Transitory Hearing Losses OR "Voice Quality" OR "Voice Qualities" OR "Breathing Exercises" OR "Obstructive Sleep Apnea" OR "Mouth Breathing" OR "Mouth Breathings" OR "Nasal Blockage" OR "Nasal Airway Obstruction" OR "Conductive Hearing Loss" OR "Dental Occlusion" OR "Canine Guidance" OR "Occlusal Guidance" OR "Communication" OR "Interdisciplinary Communication" OR "Multidisciplinary Communication" OR "Diaphragm" OR "Diaphragms" OR "Respiratory Diaphragm" OR "Rehabilitation of Speech and Language Disorders" OR "Language and Speech Disorder Rehabilitation" OR "Language and Speech Disorder Rehabilitation" OR "Speech and Language Disorder Rehabilitation" OR "Oral Myotherapy" OR "Orofacial Myology")	530	15	6	6
Lilacs	("Cystic Fibrosis" OR Mucoviscidosis OR "Pulmonary Cystic Fibrosis" OR "Fibrose Cística" OR "Fibrosis Quística" OR "Mucoviscidose") AND ("Stomatognathic System" OR "Mastication" OR "Masticación" OR "Mastigação" OR "Sleep" OR "Sleep Disorders" OR "Sono" OR "Sueño" OR "Deglutition" OR "Deglución" OR "Deglutição" OR "Swallowing" OR	27	2	2	2

	"Deglutition Disorders" OR "Swallowing Disorder" OR "Transtornos de Deglutição" OR "Trastornos de Deglución" OR "Dysphagia" OR "Oropharyngeal Dysphagia" OR "Hearing Loss" OR "Hypoacusis" OR "Hypoacuses" OR "Pérdida Auditiva" OR "Perda auditiva" OR "Hearing Impairment" OR "Voice Quality" OR "Qualidade da Voz" OR "Calidad de la Voz" OR "Voice" OR "Voz" OR "Sleep Apnea, Obstructive" OR "Apnea Obstrutiva do Sono" OR "Apnea Obstrutiva del Sueño" OR "Mouth Breathing" OR "Respiração Bucal" OR "Respiración por la Boca" OR "Speech-Language Pathology" OR "Fonoaudiologia" OR "Fonoaudiología")				
Total		1566	97	39	30

Source: Prepared by the authors (2023)

A total of 39 articles were read in full, 9 of which were excluded because they did not contain SLP data. Therefore, 30 articles were selected for data collection and make up table 2, in which all of them are related to areas or alterations in which the SLP service may be important.

Most of the articles were published after 2010, showing that they are recent studies that have been increasing in recent years (McCormick, Cho *et al.*; Nelson, Karempelis *et al.*; Jankelowitz, Reid *et al.*, 2005; Martins, Camargos *et al.*, 2010; Sabati, Kempainen *et al.*, 2010; Ramos, Salles *et al.*, 2011; Fauroux, Pepin *et al.*, 2012; Perin, Fagondes *et al.*, 2012; Spicuzza, Sciuto *et al.*, 2012; Hayes, Daniels *et al.*, 2014; Lourenço, Costa *et al.*, 2014; Dziekiewicz, Banaszkiwicz *et al.*, 2015; Geyer, Barreto *et al.*, 2015; Veronezi, Carvalho *et al.*, 2015; Doumit, Belessis *et al.*, 2016; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Isaiah, Daher *et al.*, 2019; Vijayasingam, Frost *et al.*, 2020). In table 2, it is possible to observe more detailed information collected from the articles.

Table 2- Data on authorship, year, country, sample, evaluation method, results and conclusion.

Author, year, country	Type of study	Sample	Instruments applied to measure speech-language alterations	Instrument used to diagnose CF	Main Results	Conclusion
Forcucci, 1972 (Forcucci e Stark, 1972) USA	Quantitative study	31 Individuals Mean age: 9.2 years	-Audiometry - Otoscopy - Utah Language Development Test - Peabody Picture Vocabulary Test - Form A - Templin-Darley's 50 Item Screening Articulation Test	Not informed	12 subjects (39%) had unilateral or bilateral hearing loss, 11 had conductive type and 1 had severe bilateral sensorineural loss. 6 (32%) out of the 19 subjects with normal hearing had conductive alterations. 7 (22%) out of the 31 subjects had low scores on the speech-language tests.	Periodic audiological assessment can help detect middle ear alterations in children with CF and mild speech impairment. The importance of early diagnosis was highlighted.
Stavroulaki, 2002 (Stavroulaki, Vossinakis <i>et al.</i> , 2002) Greece	Prospective study control-case.	31 Individuals (12 with CF using gentamicin, 8 with CF, 11 without CF) 6-13 years	- Otoscopy - Immittance audiometry - Tonal Audiometry - Evoked OAE.	Sweat test	For the group that used gentamicin, no significant change in threshold was observed. Patients in the gentamicin group with CF had decreased emission levels	Aminoglycosides is a drug used to control infections in CF, it may be linked to decreased auditory function and suggests that evoked OAE should be included in the routine for monitoring the hearing of patients with CF.
Piltcher, 2003 (Piltcher, Teixeira <i>et al.</i> , 2003) Brazil	Review of medical records, retrospective study	42 Individuals Mean age: 7.87 years	- Audiometry - Tympanometry	Not informed	12 (28%) individuals had hearing loss. Acoustic reflex was absent in 36%. Tympanometry performed by 34 patients, 91% curve A and 8% curve B. 28.56% had sensorineural hearing loss.	It shows the lack of attention to hearing health in CF patients, which is important due to the use of medications that can cause hearing loss.
Mulheran,	Randomized	244 Individuals	- Audiological screening by questionnaire	Positive sweat test or genotyping	Considering the 244 patients who started the study, 168 had sufficient	The study concluded that the use of tobramycin at

2006 (Mulheran, Hyman-Taylor <i>et al.</i> , 2006) UK	controlled trial	> 5 years	- Otoscopy - Tympanometry - Tonal Audiometry		standard PTA data sets before and after. During the research, 13 patients were removed from the study because they already had preexisting hearing loss. In the follow-up performed at 6 and 8 weeks, no audiological changes were observed.	different dosages (once a day and 3 times a day) does not indicate cochleotoxicity, an effect that would cause hearing loss. However, the study does not provide a basis for the use of the drug in the long term or for repeated therapies, requiring a long-term study with routine monitoring of audiometry.
Cheng, 2009 (Cheng, Johnston <i>et al.</i> , 2009) USA	Review of medical records, retrospective study	171 Individuals 10.9 and 15.0 years	- Audiometry - Tympanometry	Not informed	14.0% of the individuals had SNHL 2.0% of the individuals had severe bilateral tinnitus without SNHL Individuals who have received several courses of aminoglycosides are at particularly high risk of developing SNHL	This study shows very important aspects to be taken into account, such as the high risk of pediatric patients developing SNHL and the need for all CF patients to undergo routine and longitudinal audiological assessments.
Martins, 2010 (Martins, Camargos <i>et al.</i> , 2010) Brazil	Descriptive study with a control group.	120 Individuals Mean age: 8.5 years	- Standardized questionnaires - Audiometry - Tympanometry	Antibiotics and AGS used in patients for diagnosis.	4 subjects had hearing loss by the tritone average (3 moderate and 1 severe loss) 10 subjects presented loss at high frequencies (8 and 12 KHz)	The tests performed to measure the speech-language disorders in patients with CF indicate that there is a high prevalence of hearing loss in these patients, which makes them a high-risk group that requires periodic evaluations by otorhinolaryngologists.

Geyer, 2015 (Geyer, Barreto <i>et al.</i> , 2015) Brazil	Observational and cross-sectional study	39 Individuals Between 7 and 20 years	- Audiometry - Immittance audiometry - Distortion Product Otoacoustic Emissions.	Not informed	The auditory thresholds of the right and left ears of the patients of two groups were analyzed: SG (study group) and CG (control group). The SG presented significantly higher thresholds at 250Hz, 1000Hz, 8000Hz, 9000Hz, 10,000Hz, 12,500Hz and 16,000Hz. SG showed a greater change in DPOAE when compared to CG.	A significant number of patients with CF presented alterations in High Frequency Audiometry and Otoacoustic Emissions, impairing the child's language development.
Handelsman, 2017 (Handelsman, Nasr <i>et al.</i> , 2017) USA	Observational and cross-sectional	71 Individuals > 8 years	- Audiometry - Immittance audiometry - Vestibular assessment: DVA, VNG, sinusoidal and rotational step tests.	Not informed	- 38% had evidence of impairment of the non-lateralized peripheral vestibular system - 11% had significant unilateral vestibular loss - 30% had evidence of bilateral vestibular system paresis - 79% had vestibular system dysfunction - 15% did not have vestibular system impact based on test results - 23% had hearing loss.	Patients with CF are constantly undergoing treatments with AGS, much is said about the risk of hearing loss in these patients. This study shows that in addition to the risk of hearing loss, there is also the risk of vestibular ototoxicity, which can generate vestibular dysfunction and its disabling symptoms.
Garinis AC, 2017 (Garinis, Cross <i>et al.</i> , 2017) USA	Observational and cross-sectional	81 Individuals Mean age: 26 years	- Audiometry - Immittance audiometry - Otoscopy	Not informed	- 36 (44%) subjects had normal hearing in both ears - 45 (56%) subjects had SNHL in one or more audiometric frequencies in at least one ear Participants with SNHL were older (mean age 26 years) than those with normal hearing (mean age 21 years)	The study shows that there was an effect of cumulative IV-AG dosage on SNHL of patients with CF, with greater changes in older individuals. This suggests the need for a cochleotoxicity monitoring program in patients with

					The group with higher or cumulative medication dosage, such as tobramycin, was associated with hearing loss.	CF, being necessary to include routine audiological exams in these patients to prevent and mitigate the effects of hearing loss.
Garinis, 2018 (Garinis, Keefe <i>et al.</i> , 2018) USA	Longitudinal Study	128 Subjects 91 with CF Mean age: 25 years 37 without CF Mean age: 34 years	- Audiometry - Tympanometry - Evoked OAE - Broadband search system	Not informed	48% had no hearing loss ( $\leq 20$ dB HL) in the entire frequency range from 0.25 to 16kHz. 52% had sensorineural hearing loss in at least one frequency ( $> 20$ dB HL) in this same frequency range.	The study shows the importance of using EOAT for monitoring ototoxicity in patients with CF. The broadband EPOAT test was used, showing to be promising for the identification of hearing loss.
Kreicher, 2017 (Kreicher, Bauschard <i>et al.</i> , 2018) USA	Retro-spective analysis of the database	217 Individuals Mean age: 8.3 years	- Audiometry	Patients diagnosed in the database with the ICD-9 coding	69 (31.8%) individuals had hearing loss in at least one ear. 62.3% of individuals with hearing loss had bilateral hearing loss and 37.7% had unilateral hearing loss. Distribution of type of hearing loss for 432 ears: 74.1% without hearing loss 5.3% Conductive hearing loss 2.3% Sensorineural hearing loss 5.1% mixed 13.2% undefined 64 ears had mild hearing loss 2.5% of the ears had moderate or worse hearing loss	Based on the data found, the study suggests that children with chronic sinusitis may have a lower risk of inflammatory middle ear disease and subsequent hearing loss than children with less severe sinusitis. These factors added to CF complications should be monitored frequently. The study suggests that all children with CF undergo periodic follow-ups to identify hearing loss.

Vijayasingam, 2020 (Vijayasingam, Frost <i>et al.</i> , 2020) UK	Integrated monitoring program	126 Individuals > 18 years	<ul style="list-style-type: none"> <li>- Web-based hearing test</li> <li>- Audiometry</li> <li>- Tablet-based audiometry</li> <li>- Otoscopy</li> </ul>	Genetic test	<ul style="list-style-type: none"> <li>- 45% had hearing loss in any frequency band tested with in-cabin audiometry.</li> <li>- Predominantly sensorineural hearing loss with 48% of the sample results.</li> </ul>	The study highlights hearing loss possibly caused by use of antimicrobials, generating ototoxic impact. Study data point to the importance of ototoxicity monitoring in chronic lung disease.
Lourenço, 2014 (Lourenço, Costa <i>et al.</i> , 2014)  USA	Systematic investigation	64 Individuals (41 without CF and 23 with CF) Age: 10-30 years	<ul style="list-style-type: none"> <li>- Vocal assessment: Maximum phonation time</li> <li>- Praat 5.1.36 software was used</li> <li>- GRBASI scale</li> </ul>	Sweat test.	<p>In the control group, the recordings showed clear characteristics of a euphonic voice.</p> <p>In the CF group, the recordings showed remarkable aperiodicity and reduced amplitude, high noise levels and loss of high frequency harmonics compared to controls.</p> <ul style="list-style-type: none"> <li>- significant decrease in intensity</li> <li>- increase in jitter</li> <li>- increase in shimmer</li> <li>- Much lower noise/harmonic ratio compared to controls</li> <li>- According to the GRBASI scale, CF patients showed clear signs of dysphonia.</li> </ul>	The study shows that patients with CF have significant changes in voice parameters, and the results are more noticeable in women, with a marked reduction in both HNR and overall subjective voice quality. The results suggest that these patients may benefit from voice therapy.
Milross, 2001 (Milross, Piper <i>et al.</i> , 2001) Australia	Cross-sectional analysis	32 Individuals Mean age: 8 and 27 years	<ul style="list-style-type: none"> <li>- Pittsburgh Sleep Quality Index (PSQI)</li> <li>- Polysomnography</li> </ul>	Not informed	26% of subjects had nocturnal desaturation, which they defined as 90% Spo2 for 5% of the night.	The study shows that muscle pressure on lung capacity can be considered a preventive contribution to sleep-related desaturation.

Naqvi, 2007 (Naqvi, Sotelo <i>et al.</i> , 2008) USA	Prospective study.	24 Individuals < 18 years	- Questionnaire answered by patients and their parents to assess sleep quality with additional questions about sleep, disturbances and daytime function. - Polysomnography	Not informed	43.5% reported problems falling asleep 39.1% reported problem staying asleep 30.4% snored at night 73.9% reported daytime sleepiness. Through polysomnography, it was possible to point out that children and adolescents with CF had a significant decrease in SE, prolonged REM latency and reduction in the percentage of REM.	The study shows that children and adolescents with CF have frequent sleep complaints and significant changes in sleep architecture, and that sleep disturbance in children and adolescents with CF can impact quality of life and clinical outcomes.
Ramos 2009 (Ramos, Salles <i>et al.</i> , 2009) Brazil	Observational cross-sectional study	63 Individuals Mean age: 2 and 14 years	- Polysomnography - Examination of the oral cavity - Sleep questionnaire	The diagnosis of CF was performed according to the standard criteria of the Cystic Fibrosis Foundation	OSAS identified in 35 (55.6%) patients. The OSAS group had an upper airway overbite > 2 mm. A total of 9 (25.7%) patients had moderate or severe OSAS (AI > 5)	The study suggests that deformities, craniofacial alterations and chronic rhinosinusitis, when related to lung diseases, can cause OSAS.
Perin 2012 (Perin, Fagondes <i>et al.</i> , 2012) Brazil	Cross-sectional study	76 Individuals (51 with CF and 25 without CF) Mean age: > 16 years	- Polysomnography - Forced spirometry - Pittsburgh Sleep Quality Index (PSQI) - Epworth Sleepiness Scale	Chloride sweat test	Sleep desaturation was more common in the CF group. Only two patients with CF (3.9%) had OSAS. 2.7% with SpO <sub>2</sub> ≥ 94% had sleep desaturation.	The study shows that CF patients end up having poorer sleep quality and very small changes in sleep architecture, which is not directly associated with obstructive sleep events.
Fauroux	Multicenter Study	80 Individuals	- Pittsburgh Sleep Quality Index - Epworth Sleepiness Scale	Chloride sweat test	Subjects reported poor sleep quality.	This study reports that most patients had poor sleep quality, which is

2012 (Fauroux, Pepin <i>et al.</i> , 2012) France		Mean age: 24 ± 10 years	- Visual analogue sleep scale (S-VAS) - Bond and Lader Visual analogue scale (VAS) - Actigraphy - Polysomnography		11% of patients had an average of 30% of the night with SpO <sub>2</sub> < 90%. 47% of subjects had nocturnal hypercapnia.	common in patients with CF, and recommends systematic screening for hypoxemia and nocturnal hypercapnia.
Spicuzza, 2012 (Spicuzza, Sciuto <i>et al.</i> , 2012) Spain	Control case	58 Individuals (40 with CF and 18 without CF) Mean age: 6 months and 11 years	- Standard spirometry - Polysomnography	Not informed	28 (70%) subjects with CF had mild to moderate obstructive sleep apnea 10 (26%) subjects with CF had adenotonsillar hypertrophy 36% had chronic rhinosinusitis. 5% had both conditions.	The study considered the existence of an early occurrence of OSA in infants and children, with the possibility of a close relationship between the occurrence of structural alteration of the oropharynx, chronic rhinosinusitis and OSA.
Jankelowitz, 2013 (Jankelowitz, Reid <i>et al.</i> , 2005) USA	Cross-sectional observational study	20 Individuals Mean age: 18 years	- Pittsburgh Sleep Quality Index - Epworth Sleepiness Scale - Medical summary form of 36 items - SF-36 - Spirometry	- Chloride sweat test - Presence of two disease-causing mutations.	Mean PSQI for patients with CF was 6.45-3.31, which was significantly higher than for control with 4.55-2.21. The mean ESS scores for the CF and control groups were 6.75 3.32 and 5.72 3.63, respectively.	The study shows that the PSQI can be useful in detecting CF patients with poor sleep quality, and that stable CF patients have sleep disruption and this may be in part related to the severity of the lung disease.
Hayes, 2014 (Hayes, Daniels <i>et al.</i> , 2014) USA	Observational and retrospective study	18 Individuals Age: 33.2 ± 2.2 years	- Polysomnography	Chloride sweat test Genetic test	56% (10/18) of patients had PH 94% (17/18) of patients completed the PSG and RHC	The study shows that patients with CF have severe changes in gas exchange when related to PH. Further studies are necessary.

Veronezi, 2015 (Veronezi, Carvalho <i>et al.</i> , 2015) Brazil	Cross-sectional study	34 Individuals Mean age: 15.9 ± 7.0 years	- SpO <sup>2</sup> measurement - Epworth Sleepiness Scale. - Portable Polysomnography - Spirometry	At least 2 sweat chloride measurements and/or identification of two CF-related mutations.	- Mean FEV was 71± 31% - Mean SpO <sup>2</sup> was 95.9 ± 1.9% - 26.47%, all children, presented malnutrition. 6 were at nutritional risk and 9 were malnourished	The study shows that CF patients with nutritional and SpO <sup>2</sup> alterations in wakefulness and room air are more likely to have sleep apnea.
Urruti 2018 (Íscar-Urrutia, Madrid-Carbajal <i>et al.</i> , 2018) Spain	Prospective descriptive cross-sectional study	23 Individuals Age: 32 ± 18 years	- CFQR 14+ Questionnaire - Pittsburgh Sleep Quality Index (PSQI) - Polysomnography	Genetic test	- Lower QoL - 57% were classified as bad sleepers - 30% had a sleep latency of 30 minutes - 43% had sleep efficiency < 85% - 35% of 19 patients had sleep disturbance more than 1x per week. - 63% of 19 patients had intra-sleep wakefulness greater than 20 minutes - 68.4% of 19 patients have disturbed REM sleep - 61% of 19 patients present the ratio between TST and bedtime < 90% - 16% of 19 patients had nocturnal hypoxia Therefore, significant changes are observed both in the results obtained by the questionnaire and by the Polysomnography.	The study shows that, according to the results of objective and subjective tests applied, it decreases in adult patients. Changes in pulmonary structures and gas exchange during sleep are risk factors for decreased sleep quality and should be observed and monitored in patients with CF.
Isaiah AMD,	Retro-spective of	35 Individuals	- Polysomnography	Genetic test Sweat electrolyte test	- 51% had OSA - 43% had sleep hypoxemia	The study showed that half of the children have OSA. Supporting pulmonary

2019 (Isaiah, Daher <i>et al.</i> , 2019) USA	medical records	Mean age: 11.6 years			-43% were on oxygen supplementation. - Children with hypoxemia had lower pFEV1 than children without hypoxemia: (44.1% [37.8, 50.5] vs. 73.2% [63.4, 82.9].	function testing, using pFEV1 as a predictor of sleep hypoxemia.
Ramos, 2011 (Ramos, Salles <i>et al.</i> , 2011) Brazil	Sequential sampling, non-probabilistic	67 Individuals Mean age: 8 years	- Questionnaires on sleep disorders - Polysomnography	Not informed	The mean and standard deviation of TTS (minutes) and sleep efficiency (%) of the patients were 379±60 and 81±11, respectively. A total of 38 (56.7%) patients had AI ≥ 1 event per hour of sleep, with 7 (18%) diagnosed with moderate OSAS, 2 with severe OSAS, and 29 (43.3%) without OSAS.	The study concludes that patients with CF have frequent sleep-related complaints, changes in sleep architecture and a high prevalence of SDB.
Sabati 2010 (Sabati, Kempainen <i>et al.</i> , 2010) USA	Cross-sectional study with adults	201 Individuals 18 years	- Spirometry	Not informed	24% of patients had frequent symptoms, 39% had occasional symptoms, the most common being heartburn, acid regurgitation and dysphagia, and 18% GER symptoms, all of which contributing to the worsening of the respiratory condition.	This study shows the characteristics of GER and suggests that the mechanism and treatment of GER symptoms in patients with CF require further investigation.
Dziekiewicz, 2015 (Dziekiewicz, Banaszkiwicz <i>et al.</i> , 2015) Poland	Multicenter prospective study	44 Individuals Mean age: 10.4 3.6, range 3.0-17.8 years	- Not informed	Not informed	GER was diagnosed in 54.5% (24/44) of patients according to the main criteria and in 70.5% (31/44) and 50.0% (22/44) when considering additional criteria. A total of 1,585 episodes of gastroesophageal	Through this study it was possible to confirm a very high incidence of GER in children with CF. It would be interesting to evaluate the effectiveness of the treatment of these patients.

					reflux were detected by MII-pH (median 35.0; 20.0–46.3).	
Doumit, 2015 (Doumit, Belessis <i>et al.</i> , 2016) Australia	Quantitative study	39 Individuals < 4 years	- No report of speech therapy procedures.	- Neonatal screening - Sweat test or genetic mutation analysis.	We used 65 paired samples from 39 children evaluated, 18 of which were sampled on one occasion, 16 were sampled on two occasions, and 5 were sampled on three occasions. Oropharyngeal suction sample.	The study shows that the level of distress during oropharyngeal suction (OPS) was generally high, and assessing this distress during OPS collection in children with CF is important to reduce the feeling of distress.
Nelson 2017 (Nelson, Karempelis <i>et al.</i> ) USA	Prospective controlled study	18 Individuals: 12 patients with CF and 6 controls Mean age: 28.9 years	- Visualization of mucosa with otoscope - Examination of images	Not informed	A total of 11 patients with CF had nasal fluid aspiration into the lungs while lying down. The control group had no incidence of aspiration.	The study shows that mucus aspiration occurs in patients with CF and brings previous studies to prove this hypothesis.
McCormick 2018 (McCormick, Cho <i>et al.</i> ) USA	Multicenter prospective longitudinal cohort research	129 Individuals > 6 years	- SNOT-20 Questionnaire	Not informed	Results found at baseline (mean $\pm$ SD) - Ear fullness - Dizziness - Facial pain / pressure - Difficulty falling asleep - Waking up at night - Lack of good sleep - Reduced concentration	The study measures the treatment performed by the drug ivacaftor, and the questionnaire applied shows significant speech-language disorders. The study concludes that the use of the drug offers benefits to the CF population.

Table caption: F: Female, M: Male, CF: Cystic Fibrosis, PTA: Pure-Tone Audiometry, HFPTA: High Frequency Pure-Tone Audiometry, AGS: Aminoglycosides, DPOAE: Distortion Product Otoacoustic Emissions, OPS: Oropharyngeal Suction Distress, GER: Gastroesophageal reflux, TTS: Total sleep time, AI: obstructive apnea index, SDB: Sleep-disordered breathing, OSA: Obstructive sleep apnea, OSAS: Obstructive sleep apnea syndrome, AHI: Apnea-hypopnea index, PH: Pulmonary hypertension, PSQI: Pittsburgh Sleep Quality Index, ESS: Epworth Sleepiness Scale.

Source: Prepared by the authors (2023)

There was a greater concentration of articles related to audiology and sleep, the others were from different areas, but with a small number of results. The areas found show symptoms that occur due to functional and structural changes caused by CF and medication use.

### 3.1 Sleep

To evaluate the sleep quality of study participants, the polysomnography (Milross, Piper *et al.*, 2001; Naqvi, Sotelo *et al.*, 2008; Ramos, Salles *et al.*, 2009; Ramos, Salles *et al.*, 2011; Fauroux, Pepin *et al.*, 2012; Perin, Fagondes *et al.*, 2012; Spicuzza, Sciuto *et al.*, 2012; Hayes, Daniels *et al.*, 2014; Veronezi, Carvalho *et al.*, 2015; Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018; Isaiah, Daher *et al.*, 2019), and questionnaires such as the Pittsburgh Sleep Quality Index and the Epworth Sleepiness Scale (Milross, Piper *et al.*, 2001; Jankelowitz, Reid *et al.*, 2005; Fauroux, Pepin *et al.*, 2012; Perin, Fagondes *et al.*, 2012; Veronezi, Carvalho *et al.*, 2015; Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018). The results showed that most individuals have a reduced quality of sleep. Polysomnography identified Obstructive Sleep Apnea, nocturnal desaturation and reduced REM latency. Questionnaires showed difficulty falling asleep, difficulty staying asleep, and reduced sleep efficiency.

### 3.2 Audiology

To measure audiological alterations, pure tone and vocal audiometry, transient otoacoustic emissions and distortion product otoacoustic emissions were used. Most studies showed mild and moderate sensorineural hearing loss, especially in high frequencies (Forcucci e Stark, 1972; Stavroulaki, Vossinakis *et al.*, 2002; Piltcher, Teixeira *et al.*, 2003; Mulheran, Hyman-Taylor *et al.*, 2006; Cheng, Johnston *et al.*, 2009; Martins, Camargos *et al.*, 2010; Geyer, Barreto *et al.*, 2015; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Garinis, Keefe *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Vijayasingam, Frost *et al.*, 2020).

### 3.3 Voice

To assess the voice quality of individuals with CF, a study used vocal assessments: Maximum phonation time, the Praat software and the GRBAS scale, showing that there

is a relationship between CF and vocal alterations such as reduced voice amplitude (Lourenço, Costa *et al.*, 2014).

### **3.4 Dysphagia**

One study used spirometry, which made it possible to correlate dysphagia as a consequence of gastroesophageal reflux, showing the importance of a speech-language assessment for a better conduct (Sabati, Kempainen *et al.*, 2010).

The other study used, as an evaluation method, the visualization of the mucosa with an otoscope to identify possible aspirations, in which it was possible to observe aspirations (Nelson, Karempelis *et al.*).

### **3.5 Orofacial Mobility**

No test with speech-language data was used, but since suction is one of the functions of the stomatognathic system in which the SLP is responsible for enabling and rehabilitating, referral should be made (Doumit, Belessis *et al.*, 2016).

### **3.6 Articles Reporting Changes Related to More than One Area of SLP**

One article was found measuring the quality of life of patients with CF, in which the SNOT-20 questionnaire was used and showed results such as facial pain, ear fullness and difficulty sleeping (McCormick, Cho *et al.*).

Another article relates quality of life to the incidence of gastroesophageal reflux in individuals with CF (Dziekiewicz, Banasziewicz *et al.*, 2015).

## **4. DISCUSSION**

In this scoping review, professionals are described in the literature as fundamental to compose the care team for patients with CF, including: pediatricians, pulmonologists, gastroenterologists, physiotherapists, nutritionists, nurses, psychologists, pharmacists and social workers (Athanzio, Silva *et al.*, 2017). The discreet inclusion of the speech-language therapist in the interdisciplinary teams of care for patients with CF was observed.

SLP is responsible for the functions and structures of the stomatognathic system, so the SLP contribution in the multidisciplinary treatment of CF is important for the

quality of life of these patients through the improvement of orofacial structures and functions, as well as quality of sleep and hearing.

Considering the 30 articles selected, 46.66% were from the USA (McCormick, Cho *et al.*; Nelson, Karempelis *et al.*; Forcucci e Stark, 1972; Jankelowitz, Reid *et al.*, 2005; Naqvi, Sotelo *et al.*, 2008; Cheng, Johnston *et al.*, 2009; Sabati, Kempainen *et al.*, 2010; Hayes, Daniels *et al.*, 2014; Lourenço, Costa *et al.*, 2014; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Kreicher, Bauschard *et al.*, 2018; Isaiah, Daher *et al.*, 2019) published in 1972, and from 2007 to 2019. This demonstrates a considerable incidence of CF, in addition to good investments in scientific research, which may explain the amount of research carried out in the country. According to the literature, 30,000 children and adults are affected, and this frequency is 1 in every 3,500 individuals in the USA, putting the country in 12th place according to data from the World Health Organization (De Cássia Firmida e Lopes, 2011).

Several types of studies were found, such as: descriptive studies to determine the distribution of diseases or conditions related to the health of the patients involved; cross-sectional study to relate the time of evaluation and manifestation of the disease and its risk factors; longitudinal and prospective study to assess the cause and effects caused in patients; randomized and controlled study to collect information and test the effectiveness of therapeutic approaches in patients; non-probabilistic sequential sampling studies, in which the collection is based on previously defined criteria and quantitative and qualitative studies to gather data and measure the quality of life of patients with CF. Observational, cross-sectional, prospective and retrospective studies were found in greater quantity in the researches, accounting for 83.5% of the total (Nelson, Karempelis *et al.*; Milross, Piper *et al.*, 2001; Stavroulaki, Vossinakis *et al.*, 2002; Piltcher, Teixeira *et al.*, 2003; Jankelowitz, Reid *et al.*, 2005; Naqvi, Sotelo *et al.*, 2008; Cheng, Johnston *et al.*, 2009; Ramos, Salles *et al.*, 2009; Martins, Camargos *et al.*, 2010; Sabati, Kempainen *et al.*, 2010; Perin, Fagondes *et al.*, 2012; Hayes, Daniels *et al.*, 2014; Geyer, Barreto *et al.*, 2015; Veronezi, Carvalho *et al.*, 2015; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Isaiah, Daher *et al.*, 2019).

A total of 2,471 individuals participated in this scoping review, and only 49 of them were part of the healthy control, representing 1.98% of the total. The participating individuals were children, adolescents, adults and elderly males and females in the age

groups ranging from 2 to 87 years of age. According to De Cássia Firmida (2011) (De Cássia Firmida e Lopes, 2011), the number of adults with CF has grown a lot and proportionally more than the number of children. In 1990, about 30% of people registered with the CFF CysticFibrosis Foundation were 18 years or older. In 2009, over 47% of the registered people were adults and this number continues to grow.

Regarding diagnosis strategy for selecting individuals for research, 3.3% of the articles used the sweat test as a diagnosis (Stavroulaki, Vossinakis *et al.*, 2002; Jankelowitz, Reid *et al.*, 2005; Mulheran, Hyman-Taylor *et al.*, 2006; Fauroux, Pepin *et al.*, 2012; Perin, Fagondes *et al.*, 2012; Hayes, Daniels *et al.*, 2014; Lourenço, Costa *et al.*, 2014; Veronezi, Carvalho *et al.*, 2015; Doumit, Belessis *et al.*, 2016; Isaiah, Daher *et al.*, 2019), and 6.6% used the genetic test (Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018; Vijayasingam, Frost *et al.*, 2020). An article published in 2008 shows that the sweat test is the main test to confirm the diagnosis of CF in adults. However, according to the Brazilian guidelines for the diagnosis and treatment of CF, the identification of the pathology can be done in the newborn screening through the levels of IRT (immunoreactive trypsinogen), and confirmed with the sweat and/or genetic test (Athanzio, Silva *et al.*, 2017).

Regarding the evaluation methods used to identify speech-language disorders, it was observed that most studies used standardized tests for each respective area. Considering the 30 articles selected for this research, 12 evaluated sleep alterations (Milross, Piper *et al.*, 2001; Jankelowitz, Reid *et al.*, 2005; Naqvi, Sotelo *et al.*, 2008; Ramos, Salles *et al.*, 2009; Ramos, Salles *et al.*, 2011; Fauroux, Pepin *et al.*, 2012; Perin, Fagondes *et al.*, 2012; Spicuzza, Sciuto *et al.*, 2012; Hayes, Daniels *et al.*, 2014; Veronezi, Carvalho *et al.*, 2015; Íscar-Urrutia, Madrid-Carbajal *et al.*, 2018; Isaiah, Daher *et al.*, 2019) and only 1 did not use Polysomnography (Jankelowitz, Reid *et al.*, 2005). A study published in 2005 focused on methods used to diagnose sleep disorders. Among them, Polysomnography is cited as the best method, being considered the gold standard for the diagnosis of Sleep Disorders, as the test allows the polygraph recording of the electroencephalogram (EEG), electrooculogram (EOG), and electromyography (EMG) of the menton and limbs, oronasal flow measurements, thoracoabdominal movement, electrocardiogram (ECG), pulse oximetry, body position, esophageal pressure measurements, snoring and supplementary EEG leads (Togeiro e Smith, 2005).

Studies related to sleep cite desaturation/hypoxemia, with breathing being directly linked to sleep, showing that patients with CF have frequent complaints about the quality of their sleep due to desaturation during the night (Ramos, Salles *et al.*, 2011). Another study carried out using Actigraphy and Polysomnography observed that patients with CF have a higher frequency of nocturnal awakenings and longer time to fall asleep, resulting in a lower quality of sleep than the group of patients without CF (Shakkottai, O'Brien *et al.*, 2018). Studies point to the need for more routine monitoring, as sleep is a factor that directly influences the quality of life. Some articles have identified Obstructive Sleep Apnea (OSA) in some patients with CF (Ramos, Salles *et al.*, 2009; Perin, Fagundes *et al.*, 2012; Spicuzza, Sciuto *et al.*, 2012). Rosa (2010) (Spicuzza, Sciuto *et al.*, 2012) shows the relationship between SLP and Obstructive Sleep Apnea. According to the author, the speech-language therapist can be inserted as early as in the diagnosis, guidelines and treatment, helping to improve the functions and structures involved in breathing during sleep.

Other articles have shown that there is a relation between hearing loss and patients with CF, which can be explained mostly by the ototoxicity caused by the frequent use of some ototoxic drugs (Forcucci e Stark, 1972; Stavroulaki, Vossinakis *et al.*, 2002; Piltcher, Teixeira *et al.*, 2003; Mulheran, Hyman-Taylor *et al.*, 2006; Cheng, Johnston *et al.*, 2009; Martins, Camargos *et al.*, 2010; Geyer, Barreto *et al.*, 2015; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Garinis, Keefe *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Vijayasingam, Frost *et al.*, 2020). According to a study published in 2005, ototoxicity can be defined as disorders caused by drugs that compromise the auditory function and/or the peripheral vestibular system and that is characterized by a sensorineural hearing loss of more than 25 db in one or more frequencies in the range of 250 to 8000hz (Hyppolito e Oliveira, 2005).

Regarding hearing (Forcucci e Stark, 1972; Stavroulaki, Vossinakis *et al.*, 2002; Piltcher, Teixeira *et al.*, 2003; Mulheran, Hyman-Taylor *et al.*, 2006; Cheng, Johnston *et al.*, 2009; Martins, Camargos *et al.*, 2010; Geyer, Barreto *et al.*, 2015; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Garinis, Keefe *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Vijayasingam, Frost *et al.*, 2020), the research shows that all articles present patients with some type and degree of hearing loss or some alteration in the middle ear (Forcucci e Stark, 1972). According to a study published in 2017, CF patients without hearing complaints may have some type of loss at high frequencies. This finding can be

justified by the fact that these patients are subject to ototoxicity, assuming that audiometry at high frequencies and otoacoustic emissions can be an adequate method for diagnosis and follow-up of CF patients at risk of ototoxicity (Caumo, Geyer *et al.*, 2017). The results of the studies showed that there are significant alterations in the field of SLP in patients with CF. Studies on hearing evidence the need for a more rigorous audiological follow-up in patients with CF (Forcucci e Stark, 1972; Stavroulaki, Vossinakis *et al.*, 2002; Piltcher, Teixeira *et al.*, 2003; Mulheran, Hyman-Taylor *et al.*, 2006; Cheng, Johnston *et al.*, 2009; Martins, Camargos *et al.*, 2010; Geyer, Barreto *et al.*, 2015; Garinis, Cross *et al.*, 2017; Handelsman, Nasr *et al.*, 2017; Garinis, Keefe *et al.*, 2018; Kreicher, Bauschard *et al.*, 2018; Vijayasingam, Frost *et al.*, 2020). A study published in 2006 focuses on the importance of audiological monitoring for patients who use ototoxic drugs. Technological advances make this monitoring possible more quickly and effectively, being able to identify hearing loss early, reducing or interrupting the damage. (Jacob, Aguiar *et al.*, 2006).

CF is a disease that requires constant treatment and monitoring. Therefore, these patients undergo daily examinations, use of antibiotics, therapies and hospitalizations. Most of these treatments are considered vital, as without them lifespan decreases. However, when it comes to research, family members and guardians are reluctant to agree to the participation of these patients, as submitting them to more exams and more therapies in addition to routine ones can become stressful, which may justify the difficulty in collecting data and conducting research in several areas, especially in the speech-language field. Even today, SLP treatment is seen by many people as non-vital, because the lack of a SLP intervention will not generate immediate death. However, some studies confirm that by improving the quality of life of CF patients, their longevity is extended. Based on this observation, it is important to jeopardize more randomized clinical trials, deepening the speech therapy therapeutic actions, being this the most important limitation (HELRIGLE, PEREIRA, LEMOS, 2014).

As it is a new and little explored area of activity, CF is not a pathology commonly addressed in the undergraduate courses, so many professionals do not have theoretical and practical knowledge for the best management of these patients. There is a need for the academy to build protocols of SLP evaluation, including all areas of interface, based on high rated research studies as randomized clinical trials. The speech-language therapist

must be included in the multidisciplinary team for the treatment of CF, regarding a longer life expectancy with improved quality of life.

## 5 CONCLUSION

The scoping review showed the alterations related to SLP that impact the lives of people with CF, among them, decreased sleep quality, sleep breathing disorders as obstructive sleep apnea, sensorineural hearing loss, reduced vocal range, dysphagia, difficulty in suction, facial pain and bronchoaspiration. Most of the studies were limited to the SLP evaluation, confirming an existence of worsed functions of breathing, mastication and deglutition, and on speech and voice, but unfortunately there are missing protocols for interventions. It is noteworthy that the publications in the area are still scarce, requiring further research, to enhance the strength of the effect of the intervention. Based on the data provided by this review, SLP is of paramount importance in the routine monitoring of these patients, as part of the multidisciplinary team for the diagnosis, guidance and treatment of the changes involved in hearing, sleep, swallowing, sucking, dysphagia, voice and quality of life.

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