

University of Northern Colorado

## Scholarship & Creative Works @ Digital UNC

---

Capstones & Scholarly Projects

Student Research

---

5-2020

### AUDIOLOGISTS' KNOWLEDGE, ATTITUDES, AND PRACTICES REGARDING PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND HEARING LOSS

Carla White  
carla.white1138@gmail.com

Follow this and additional works at: <https://digscholarship.unco.edu/capstones>

---

#### Recommended Citation

White, Carla, "AUDIOLOGISTS' KNOWLEDGE, ATTITUDES, AND PRACTICES REGARDING PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND HEARING LOSS" (2020). *Capstones & Scholarly Projects*. 73.  
<https://digscholarship.unco.edu/capstones/73>

This Dissertation/Thesis is brought to you for free and open access by the Student Research at Scholarship & Creative Works @ Digital UNC. It has been accepted for inclusion in Capstones & Scholarly Projects by an authorized administrator of Scholarship & Creative Works @ Digital UNC. For more information, please contact [Jane.Monson@unco.edu](mailto:Jane.Monson@unco.edu).

UNIVERSITY OF NORTHERN COLORADO

Greeley, Colorado

The Graduate School

AUDIOLOGISTS' KNOWLEDGE, ATTITUDES, AND  
PRACTICES REGARDING PATIENTS WITH  
SYSTEMIC LUPUS ERYTHEMATOSUS  
AND HEARING LOSS

A Capstone Research Project Submitted in Partial Fulfillment  
of the Requirements for the Degree of  
Doctor of Audiology

Carla M. White

College of Natural and Health Sciences  
School of Audiology and Speech-Language Sciences  
Audiology

May 2020

This Capstone Research Project by: Carla M. White

Entitled: *Audiologists' Knowledge, Attitudes, and Practices Regarding Patients with Systemic Lupus Erythematosus and Hearing Loss*

has been approved as meeting the requirements for the Degree of Doctor of Audiology in College of Natural and Health Sciences in School of Audiology and Speech-Language Sciences, Program of Audiology

Accepted by the Capstone Research Committee

---

Deanna K. Meinke, Ph.D., Research Advisor

---

Kathryn Bright, Ph.D., Committee Member

---

Kimberly Murza, Ph.D., Committee Member

Accepted by the Graduate School

---

Cindy Wesley  
Interim Associate Provost and Dean  
Graduate School and International Admissions

## ABSTRACT

White, Carla M. *Audiologists' Knowledge, Attitudes, and Practices Regarding Patients with Systemic Lupus Erythematosus and Hearing Loss*. Unpublished Doctor of Audiology capstone research project, University of Northern Colorado, 2020.

Patients diagnosed with systemic lupus erythematosus (SLE) have a moderate to high risk of developing a sensorineural hearing loss. The purpose of this study was to identify the knowledge, the degree of confidence, and the clinical practices of audiologists and graduate students when providing audiological care to this population. An opportunity for the respondents to share clinical experiences and knowledge was also provided.

A sample of 77 respondents, 24 licensed audiologists and 53 graduate students, represented that audiologists are generally aware of the disease and that SLE can cause sensorineural hearing loss. Graduate students were more likely to learn about SLE from personal relationships, while licensed audiologists first learned of SLE from another health care professional. Outcomes also revealed that additional resources, such as a clinical protocol and a list of SLE support groups, are needed and have been requested by both respondent groups.

Both licensed audiologists and graduate students are knowledgeable with SLE but are currently interested in clinical resources. There is an opportunity for additional research to create a more formal and finite protocol to be utilized when an audiologist

sees a patient with SLE. Further research should identify the appropriate actions an audiologist must complete in order to ensure that sufficient long-term audiologic care can be provided to patients diagnosed with systemic lupus erythematosus

## ACKNOWLEDGMENTS

My endless gratitude goes to Dr. Deanna Meinke, the research advisor of this project. I cannot thank her enough for the patience and wealth of knowledge she has provided over the course of my graduate career as a professor and advisor. I would also like to acknowledge Dr. Katie Bright and Dr. Kim Murza for their recommendations and encouragement for the completion of this capstone research project.

This project was completed by the encouragement of my family, my friends, and my colleagues. My parents, Giselle White and Zach T. White Jr., have been there every step of the way of my educational career and even more so. It is these two individuals who have cheered me on to accomplish this dream, even when I was halfway across the United States.

I also want to thank the individuals who took the time out of their busy days to complete this survey. These are the people who allowed this project to come together and provide the information needed to initial further research.

## TABLE OF CONTENTS

CHAPTER	
I.	INTRODUCTION ..... 1 Statement of the Problem Rationale Research Questions
II.	REVIEW OF THE LITERATURE ..... 4 Autoimmune Diseases Systemic Lupus Erythematosus Hearing Loss Characteristics Vertigo Tinnitus Identification of Autoimmune-Related Hearing Loss Ototoxic Medications Resources and Support Groups Future Research
III.	METHODS ..... 28 Development of the Survey Instrument Participants Survey Instrument Sampling Methods Descriptive Analysis
IV.	RESULTS ..... 33 Respondent Characteristics Knowledge Outcomes Attitudes of Licensed Audiologists and Audiology Graduate Students Clinical Practice for Systemic Lupus Erythematosus Patients Open-Ended Comments Summary

CHAPTER	
V.	DISCUSSION ..... 57
	Experience of Providing Care
	Need for Clinical Resources
	Study Limitations
	Future Research
	Conclusion
	REFERENCES ..... 69
APPENDIX	
A.	Institutional Review Board Approval ..... 75
B.	Consent Form and Survey ..... 78
C.	Clinical Research Articles Resource ..... 99
D.	Survey Knowledge Section Answer Key ..... 101
E.	Systemic Lupus Erythematosus Patient Support Group Resource ..... 103
F.	Clinical Protocol Guideline ..... 105



## LIST OF TABLES

### TABLE

1.	Program Status of Graduate Student Respondents .....	35
2.	Awareness of Systemic Lupus Erythematosus (SLE) and Awareness of SLE Related Hearing Loss by Graduate Students and Audiologists .....	39
3.	Sources of Systemic Lupus Erythematosus (SLE) Information Identified by Graduate Students and Audiologists .....	41
4.	How Respondents were First Aware of the Relationship of Hearing Loss and Systemic Lupus Erythematosus (SLE) .....	42
5.	Number of Correct Responses between Graduate Students and Licensed Audiologists .....	45
6.	Tests that Should be Added to the Standard Audiometric Battery for Patients being Monitored for Systemic Lupus Erythematosus (SLE) .....	47
7.	Response Summary to Survey Question “How Often Do You Recommend Patients with Systemic Lupus Erythematosus to Have a Hearing Evaluation?” .....	51
8.	Additional Audiometric Tests Provided to Patients with Systemic Lupus Erythematosus .....	52
9.	Summary of Types of Information that Would be Useful to Graduate Students and Audiologists with Regard to Providing Audiologic Care to Patients with Systemic Lupus Erythematosus (SLE) .....	54

## LIST OF FIGURES

### FIGURE

1.	Number of Cumulative Years of Practice and Number of Years In a Medical Setting Reported by Audiologist Respondents .....	36
2.	Practice Setting of the Licensed Audiologists .....	37
3.	How Long Ago were Graduate Students and Licensed Audiologists First Aware of Systemic Lupus Erythematosus .....	43
4.	Comparison of Mean Likert Scores between Graduate Students and Licensed Audiologists .....	47
5.	Estimated Number of Patients with Systemic Lupus Erythematosus (SLE) Who have been Cared for with Licensed Audiologists and Graduate Students .....	49
6.	Summary of Referrals to Other Specialists Made by Audiology Graduate Students and Audiologists .....	52

## LIST OF ABBREVIATIONS

ABBREVIATION	MEANING
AAA	American Academy of Audiology
ABR	Auditory Brainstem Response
ANA	Antinuclear Antibodies
APD	Auditory Processing Disorder
APS	Antiphospholipid Syndrome
ART	Acoustic Reflex Threshold
BPPV	Benign Paroxysmal Positional Vertigo
CEUs	Continuing Education Units
CHL	Conductive Hearing Loss
DPOAEs	Distortion Product Otoacoustic Emissions
ECochG	Electrocochleography
EHFA	Extended High-Frequency Audiometry
ENG	Electronystagmography
HFOAEs	High-Frequency Otoacoustic Emissions
HIV	Human Immunodeficiency Virus
HL	Hearing Level
NHIS	National Health Interview Survey
OAEs	Otoacoustic Emissions
OHC	Outer Hair Cells
SLE	Systemic Lupus Erythematosus
SLEDAI	Systemic Lupus Erythematosus Disease Activity Index
SLICC	Systemic Lupus International Collaborating Clinics
SNHL	Sensorineural Hearing Loss
TEOAEs	Transient Evoked Otoacoustic Emissions
VEMP	Vestibular Evoked Myogenic Potential
VNG	Videonystagmography

## CHAPTER I

### INTRODUCTION

Systemic lupus erythematosus (SLE), in short, can be described as increased autoimmune activity. While healthy and typical autoimmune responses are designated to attack foreign bodies and viruses present in the human body, healthy tissues, organs, and other bodily systems are targeted by the overactive immune system. A number of elements need to be taken in account when determining the cause of the disease manifestation. Factors causing the presentation of SLE symptoms include genetic makeup (Akirav et al., 2011; Anderson & Su, 2011), environmental events (Davidson & Diamond, 2014), or a combination of these two factors. Gender and race are known determinants: the majority of SLE patients are female, and higher rates of African Americans are diagnosed with the disease (Dragin et al., 2017). Pubescent development has a role in disease initiation; therefore, the majority of SLE patients are age 20 or older (Bovo et al., 2006). These characteristics of SLE can make it challenging for clinicians to confirm the presence of the disease.

Varying organ involvement is reported with each SLE patient, and the auditory system is one of the vulnerable structures. Researchers have distinguished a number of attributes associated with autoimmune-related hearing loss. If the patient does have autoimmune involvement within the auditory system, a bilateral high-frequency sensorineural hearing loss (SNHL) that is symmetrical in nature may develop.

The majority of published work is quantitative in design. Researchers provide recommendations to audiologists in the counseling and care for SLE patients diagnosed with a hearing loss. For instance, these patients should pursue annual hearing evaluations in order to monitor the hearing loss. What remains to be analyzed is whether practicing audiologists are considering these recommendations. Research that is qualitatively designed may render valuable information related to how audiologists care and monitor SLE patients.

### **Statement of the Problem**

The general understanding of the connection between hearing loss and autoimmune responses related to SLE has been expanded upon through recent research. What should be answered is what are the clinical methods and protocols used by audiologists to evaluate and monitor autoimmune inner ear disease in SLE patients. Are audiologists practicing in the United States aware of the heterogeneous nature of SLE and that each SLE patient will have differing symptoms and secondary diagnoses that develop due to the autoimmune disease? Do audiologists stress the importance of annual hearing evaluations with this population due to unexpected flares or sudden rise of symptoms? What is the timeline of events for a patient needing a medical referral for SLE related hearing loss? How long is this process?

### **Rationale**

The purpose of this study was to evaluate the knowledge, attitudes, and practices of audiologists who see SLE patients for audiological evaluations. A survey was developed and was composed of sections to evaluate knowledge, attitudes, and practices. Additional demographic information was included, such as whether the audiologist was currently licensed and how many years they had been providing

audiological services. The purpose of the survey was to further inform best practices in hearing health care provided to patients with SLE.

### **Research Questions**

- Q1 What are the knowledge, attitudes, and practice patterns of currently practicing and experienced audiologists regarding patients with systemic lupus erythematosus?
- Q2 What are the knowledge, attitudes, and practice patterns of current audiology graduate students regarding patients with systemic lupus erythematosus?

## CHAPTER II

### REVIEW OF THE LITERATURE

Systemic lupus erythematosus (SLE) is one of many autoimmune diseases that have yet to be fully understood. While this disease is known for its impact on organs such as the kidneys, skin, and heart, current research has identified the involvement and pathophysiology of SLE on the auditory system. Over the past 30 years, multiple groups of researchers have recognized the role of SLE in the onset of sensorineural hearing loss (SNHL), tinnitus, and vertigo. Furthermore, the limited pharmacological options intended to suppress the autoimmune activity of lupus have been found to have ototoxic effects. The purpose of this literature review is to describe the degree and prevalence of auditory impairment in SLE patients. The objective of this research design is to identify the degree of knowledge and the therapy methods that are utilized by audiologists within the United States.

#### **Autoimmune Diseases**

Autoimmune diseases have been studied for centuries; however, this type of disease has yet to be fully understood. Due to the varying symptoms across patients, thorough evaluation is required in order to confirm an autoimmune disease diagnosis. Intensive testing includes multiple blood analyses to determine blood disorders and antibody levels of the immune system, urine analysis to decide kidney involvement, dermatological assessment to diagnose skin conditions, evaluation of reported pain and soreness in the joints, and possible neurological examinations (Al-Sukaini et al.,

2014). It is crucial that clinicians in multiple fields, including audiologists, are informed of how this disease may impact the physiological and anatomy that falls within their professional area of expertise.

Autoimmune disease is characterized as an immune response that presents as an inflammatory response in either a concentrated area or within multiple systems of the body (Davidson & Diamond, 2001). Diagnoses that fall within the category of autoimmune diseases include Wegener's granulomatosis, rheumatoid arthritis, type II diabetes mellitus, Sjogrens disease, Cogan syndrome, and Susac syndrome.

While the integrity and health of the body depend on the proper functioning of the immune system, prolonged periods of increased immune activity can be detrimental (Davidson & Diamond, 2014). As Davidson and Diamond (2001) explained, a typical healthy immune response is initiated when the presence of foreign bodies, such as an infection or bacteria, has been identified. Davidson and Diamond stated that groups of T-cells, B cells, and monocytes attack the foreign bodies and clear away the debris. In autoimmune diseases, areas of healthy tissues are damaged by groups of overactive T-cells (Davidson & Diamond, 2001). In general, autoimmune diseases are distinguished by the increased immune activity as well as damage of healthy tissues.

Genetic studies have revealed that each disorder that falls under the autoimmune disease umbrella has a genetic element (Akirav et al., 2011; Anderson & Su, 2011). Deletion of a section of the autoimmune regulator (AIRE) gene or a defect of the Fas gene can lead to the development of a form of autoimmunity (Fleisher et al., 2001; Grodzicky & Elkon, 2002). Additionally, the lack of the Foxp3 gene, a T-cell regulating gene, can result in an increased number of T-cells. In some cases, the



failure to remove dead material after an immune response, leading to an accumulation of material, causes an exaggerated immune response, a common feature of SLE (Lewis & Botto, 2006; Pettigrew et al., 2009).

Additional research has confirmed theories that autoimmunity can also be the result of environmental events (Davidson & Diamond, 2014). These alterations include gum infections (periodontal disease) and smoking (Klareskog et al., 2011) as well as ultraviolet light (Bijl & Kallenberg, 2006). Therefore, susceptibility to autoimmune diseases is attributed to genetic predispositions, environmental events, or a combination of the two (Davidson & Diamond, 2014).

The first signs of autoimmune disease development can occur at the age of 20 years or older (Bovo et al., 2006). Though uncommon, patients under the age of 20 have been diagnosed. Childhood-onset and juvenile-type SLE make up approximately 10% to 20% of the SLE population with the majority being diagnosed between 12 and 18 years of age (Silva et al., 2012). The majority of autoimmune diseases are more prevalent in females than males, a finding that is indicative of sexual hormone involvement (Davidson & Diamond, 2014). Studies have concluded that possessing two X chromosomes increases the susceptibility to autoimmune diseases (Smith-Bouvier et al., 2008). This can be attributed to the greater number of genes located on the X-chromosome that are responsible for immunological development (Smith-Bouvier et al., 2008).

Research completed by Dragin et al. (2017) reported that the expression of the AIRE gene, responsible for managing autoimmunity, is decreased in females after puberty. Further investigation suggests a possible relationship of the AIRE gene and estrogen, suggesting that as estrogen levels increase, AIRE gene expression is

suppressed (Dragin et al., 2017). Even with these recent findings, more research is required to better understand the complex relationship of gender, genetics, and autoimmune diseases.

Due to the complexity of these diseases, it is common to have a team of clinicians work together and provide therapeutic options. While treatment and care is typically provided at the discretion of a rheumatologist, patients diagnosed with autoimmune diseases are likely to be seen by hematologists, vascular care professionals, and audiologists. An overwhelming amount of evidence suggests that select autoimmune diseases negatively impact the auditory system and cause different degrees of hearing loss, affect the vestibular system, and are associated with a prevalence of tinnitus. This type of knowledge can be used to educate audiologists regarding the auditory structures that are impacted. As a result, an audiologist can select therapeutic options and additions to the audiometric test battery to provide the comprehensive medical surveillance and rehabilitative care this population requires.

### **Systemic Lupus Erythematosus**

“Lupus” is the latin term for “wolf” and was first used by Pierre Louis Alpheé Cazenave, a French physician, when he observed malar rashes in some SLE patients that resembled the shape and color of the bite of a wolf. (Chabner, 2013).

“Erythematosus,” stemming from the root word “erythema,” is defined as redness of the skin (Kim & Werth, 2014). As mentioned earlier, the malar rash, or malar erythema, is a common feature of SLE and may be referred to as a “butterfly rash” (Kim & Werth, 2014, p. 37).

## **Diagnostic Criteria**

The SLE is an autoimmune disease that is predominately diagnosed in the young (Batuecas-Caletrío et al., 2013). Features of this disease can vary extensively from patient to patient. Yu et al. (2014) published a review that discussed the most recent update to the diagnostic criteria for SLE by the Systemic Lupus International Collaborating Clinics (SLICC) in 2012. A diagnosis is determined if the individual satisfies at least four features that fall into the following categories: abnormal immunological features, hematologic disorder, renal disorders, serositis (inflammation of serous tissues that surround the heart, lungs, and abdomen), symptoms affecting the joints, and cutaneous disorders (Yu et al., 2014). According to the Rheumatology Boston weighted criteria, common features of SLE include positive antinuclear antibody (ANA) counts within the blood, cytopenia (reduced blood cell count), arthritis, malar rash across the face, and photosensitivity (Costenbader et al., 2002). These findings were based on the reported symptoms of SLE patients receiving care at Massachusetts General Hospital Rheumatology Clinic (Costenbader et al., 2002). Due to the considerable involvement of SLE throughout the body, the auditory system is at risk of a magnified inflammatory response.

## **Prevalence**

The Centers for Disease Control and Prevention have funded a number of registries, as well as longitudinal research, with the intention of identifying the prevalence of SLE in the United States in five regions: San Francisco County in California, Manhattan in New York, Fulton and DeKalb counties in Georgia, and Wayne and Washtenaw counties in Michigan (Dall’Era et al., 2017; Izmirlly et al., 2017; Lim et al., 2014; Somers et al., 2014). With these registries, the accuracy of the

outcomes is questionable and possibly underestimated due to the exclusion of patients within the Veterans Health Administration system (Izmirly et al., 2017), the limited hospitals reviewed due to other facilities that declined to participate (Izmirly et al., 2017), unidentified SLE cases (Somers et al., 2014), and the retrospective nature of the research (Dall'Era et al., 2017; Lim et al., 2014; Somers et al., 2014).

Prevalence rates varied within each region. The highest value of 84.8 for every 100,000 individuals was reported in San Francisco County between the years 2007 and 2009 (Dall'Era et al., 2017). The prevalence rates were slightly lower in New York with amounts at 62.2 for every 100,000 Manhattan citizens between 2007 and 2009 (Izmirly et al., 2017). Lim et al. (2014) reported rates in Georgia of 74.4 per 100,000 and 72.8 per 100,000 in Michigan. In all of these studies, a greater prevalence was associated with women and individuals of African American background.

Other efforts have been devoted to the evaluation of SLE prevalence in the United States. Helmick et al. (2008) used the 2003-2005 National Health Interview Survey (NHIS) and determined that between 161,000 and 322,000 adults were living with the disease at the time. Outcomes from the efforts of the Centers for Disease Control and Prevention and NHIS provide a preliminary of the number of SLE patients; however, further investigation is required to establish definite prevalence rates.

To evaluate of the prevalence of hearing loss in SLE patients, Rahne et al. (2017) compared 20 individuals with SLE to a control group that was matched for both gender and age. The SLE patients were assessed according to duration of SLE, what organs were impacted, the use of steroids over the time in which the disease had been treated, and the use of immunosuppressive medications. The hearing evaluation

completed with both participant groups included tympanometry, acoustic reflex testing, pure-tone audiometry, sentence testing with speech perception and word recognition, and distortion product otoacoustic emissions (OAEs). Pure-tone audiometric testing resulted in higher rates of SNHL associated with the SLE group compared to the control group. The results from the other tests in the audiometric test battery were not significantly different between the two groups. Rahne et al. interpreted these results to be the consequence of disease involvement affecting the stria vascularis, spiral ganglion cells, or hair cells, or the result of microinfarctions of temporal microvessels. From this research, it was estimated that there is a moderate to high association of hearing loss with SLE (Rahne et al., 2017).

### **Temporal Bone Studies**

Numerous auditory structures are impacted by SLE. Sone et al. (1999) evaluated the structural integrity of the temporal bones from seven patients, ages 14 to 76 years, diagnosed with SLE. The presence of the stria vascularis, cochlear hair cells, and ganglion cells was measured. The degree of loss of each auditory structure varied by patient with case one having little involvement while cases three and seven showed high losses of these structures from the cochlear base to the apex.

Kariya et al. (2016) examined a larger participant pool of temporal bones: 15 from SLE patients and an additional 17 bones that made up the age-matched control group (between 14 and 76 years of age). Kariya et al. concluded that the amount of stria vascularis tissue of all the cochlear turns was significantly reduced when compared to normal temporal bones ( $p < 0.05$ ). Significant findings were also present with the degree of outer hair cell loss ( $p < 0.05$ ). Inner hair cells were also impacted yet this was not statistically significant ( $p < 0.05$ ). Within this study, the most severe

case was a 22-year-old woman with complete obliteration of cochlear structures such as the organ of Corti, stria vascularis, and basilar membrane. These structures were replaced with fibrous tissue left behind by the inflammatory response. In conclusion, the invasive nature of SLE can result in the irreversible deterioration of multiple auditory structures. Therefore, it is paramount that the audiologist promptly identifies the hearing loss.

### **Hearing Loss Characteristics**

Research completed in the late 1990s to today has identified varying degrees and types of SNHL associated with SLE. Studies have been designed to recruit SLE patients that are below the age in which presbycusis becomes a possible factor for the onset of the hearing loss. When identifying the most common symptoms impacting the auditory system, the researchers reported that prevalence of hearing loss due to SLE ranged from 6% to 70% (Di Stadio & Ralli, 2017). There is little agreement across the completed research studies about the range of frequencies that are impacted by SLE.

### **Disease Duration**

Variable outcomes are also reported when disease duration is compared to the degree of the hearing loss. The purpose of the study by Maciazczyk et al. (2011) was to identify whether there was a relationship between these two factors. Thirty-seven SLE patients were matched according to age and gender with a control group. The SLE patient group was then divided into three groups according to disease duration: five years or less, six to 10 years, and over 10 years. The mean age was 42.6 years for the first group, 51.6 years for the second group, and 52.8 years for the third group. To prevent the misinterpretation of hearing loss due to presbycusis rather than SLE, each group was divided further into younger and older groups. Therefore, a total of six

groups were evaluated. Maciazczyk et al. concluded that the duration of the disease had a greater impact on audiometric thresholds than did age. This allows researchers to rule out the role of presbycusis. Consequently, audiologists should be aware that patients diagnosed with SLE early in their lives exhibit significantly decreased auditory thresholds that are unrelated to presbycusis.

### **Frequencies**

Di Stadio and Ralli (2017) completed a meta-analysis of the literature with regard to SLE and the presence of SNHL. A prevalence rate of 65% for SNHL affecting the higher frequencies, specifically 4000 and 8000 Hz was identified. Abbasi et al. (2013) identified 12 of the 45 (26.7%) participants with SLE as having bilateral high-frequency SNHL. Roverano et al. (2006) established that of 31 patients with a mean age of 35 years, 66% had a bilateral SNHL in the higher frequencies. Additionally, Maciazczyk et al. (2011) reported that 28.6% of the recruited SLE patients had a high-frequency hearing loss when a standard audiometric battery was completed. In conclusion, the onset of a high-frequency hearing loss is an apparent characteristic of SNHL in individuals diagnosed with SLE.

Di Stadio and Ralli (2017) also reported that the second-most prevalent type of SNHL (32%) occurred within the mid-frequencies, which affected 2000 and 3000 Hz. Karatas et al. (2007) confirmed that 21% of the participants had a SNHL at 500, 1000, and 2000 Hz. These outcomes support the proposition that autoimmune-related hearing loss is not solely restricted to the higher frequencies.

Low-frequency SNHL occurred with a 3% prevalence rate in the published literature (Di Stadio & Ralli, 2017). Rahne et al. (2017) evaluated 58 patients with three autoimmune disorders, one of which included SLE. Results established that SLE

patients were predisposed to the development of low-frequency SNHL. Karabulut et al. (2010) suspected that the decreased audiometric thresholds in this frequency range could be indicative of either endolymphatic or cochlear hydrops. Altogether, some SLE patients may have a SNHL affecting the frequencies between 4000 and 8000 Hz, while others have a SNHL impacting frequencies below 1000 Hz. This may be attributed to the heterogenous nature of SLE in which each SLE patient can have differing symptoms.

While results from multiple studies suggest that the SNHL is focused within a specific range of frequencies, other studies have identified SNHL across a range of frequencies. Andonopoulos et al. (1995) determined a significant decrease in auditory thresholds at 125 and 500 Hz in the study group comprised of participants between the ages 16 and 59 years. A second finding revealed a decrease in the higher frequencies was observed in the group that was referred to as the young patients and incorporated SLE patients between the ages of 16 and 29 years. Karabulut et al. (2010), after completing pure-tone audiometry, found significantly decreased auditory thresholds at 250, 500, 1000, 2000, 10,000, and 12,000 Hz. These findings indicate that both low and high frequencies could be significantly impacted. In conclusion, a comprehensive review of the literature demonstrates that high-frequency SNHLs are frequently identified. However, reduced auditory thresholds have been identified in other conventional audiometric frequency ranges.

### **Progressive Versus Static Hearing Loss**

An additional feature of SNHL in SLE patients that needs to be evaluated is the time course: progressive or static. While this information is limited in its relation



to systemic lupus, a few studies explored the longitudinal status of the hearing loss. An early study by Sperling et al. (1996) associated a slowly progressive SNHL due to the mechanisms of SLE. Cordeschi et al. (2004) had similar results with the use of transient evoked otoacoustic emissions (TEOAEs). In antithesis of these findings, Maciazczyk et al. (2011) determined that the hearing losses were static and not susceptible to further decline.

### **Sudden Sensorineural Hearing Loss**

The SNHL with a sudden onset is a possible, but uncommon, event associated with SLE. In a number of patient cases, the sudden hearing loss was the initial symptom of SLE (Green & Miller, 2001; Kastanioudakis et al., 2002). Lin et al. (2013) identified greater incidence rates of sudden SNHL in SLE patients when compared to participants without the autoimmune disease. Furthermore, SLE patients over the age of 35 years had a greater incidence of sudden SNHL (Lin et al., 2013).

Researchers have been aware of this feature of hearing loss associated with SLE since the 1990s. One theory supporting the sudden onset includes a condition that is frequently comorbid with SLE: antiphospholipid syndrome (APS). The APS is known to cause microthrombosis of the stria vascularis (Hisashi et al., 1993). A second theory states that the activity of the autoimmune antibodies can result in the diminished numbers of inner ear proteins (Suzuki et al., 1997).

An additional feature to note is whether the patient with SLE has any comorbid conditions, such as rheumatoid arthritis. Xie et al. (2019) focused on the likelihood of recovery from a sudden SNHL in patients with both SLE and rheumatoid arthritis.

Findings suggest that the prognosis of this particular set of patients was low and that

there was a low likelihood that hearing sensitivity would recover. Riera et al. (2019) identified the same outcome in patients with SLE and APS. Overall, the additional comorbidities can result in a poorer prognosis in terms of sudden SNHL.

While the incidence is not high, the sudden onset of SNHL should be recognized as a potential feature in an SLE case. In addition, if a sudden SNHL is confirmed in a patient, the audiologist and otolaryngologist must rule out the likelihood of autoimmunity as the cause of the hearing loss. Lin et al. (2013) recommended that SLE patients should have regular evaluations for the early determination of autoimmune-related sudden SNHL.

### **Bilateral Versus Unilateral Hearing Loss**

Within the literature, studies have explored whether SNHL in SLE manifests predominately as a bilateral loss or as a unilateral loss. The majority of published work has identified the prevalence of bilateral SNHL within this specific population (Abbasi et al., 2013; Kastanioudakis et al., 2002; Maciazczyk et al., 2011; Roverano et al., 2006; Sperling et al., 1996). However, a number of research studies have identified cases of both bilateral and unilateral SNHL (Gomides et al., 2007; Karatas et al., 2007). Gomides et al. (2007) identified three patients with a bilateral loss and four patients with unilateral losses, approximately 57% of the identified participants. Similar findings were found by Karatas et al. (2007); three individuals were identified with a unilateral SNHL and three other individuals with bilateral SNHL. Overall, evidence confirms that a bilateral SNHL is more likely to manifest; however, an audiologist should anticipate the possibility of identifying either a unilateral or bilateral SNHL caused by the autoimmune response related to SLE.

## **Symmetry**

The symmetry of a bilateral hearing loss attributed to SLE should be considered. Published literature has focused on the identification of the frequencies impacted and provides only a limited amount of information regarding whether the hearing losses were symmetrical or asymmetrical. Early findings from Sperling et al. (1996) concluded that asymmetric hearing losses were prevalent in lupus patients. Others have found contrasting results to Sperling et al. (1996). The 20 patients identified by Roverano et al. (2006) with a SNHL loss were all symmetrical. Additionally, both standard audiometric testing and auditory brainstem response (ABR) testing by Maciazczyk et al. (2011) identified symmetrical bilateral losses in the recruited participant group. From this analysis, it appears that there is greater prevalence of symmetrical SNHL than asymmetrical SNHL in systemic lupus patients.

## **Hearing Loss Severity**

The following parameters of hearing loss associated with SLE have been identified: frequency and symmetry. The degree or severity of disease activity is an additional feature that is utilized to anticipate a hearing loss. In the previous study, Maciazczyk et al. (2011) evaluated the patients according to the severity of renal conditions, central nervous system conditions, and antibody levels, all of which are influenced by SLE severity. An audiometric evaluation consisting of pure-tone audiometry, speech recognition testing, word recognition testing, tympanometry, and auditory brainstem response testing was completed and results were compared to those of a control group consisting of age-matched participants with normal hearing sensitivity and no health conditions. The degree of the hearing loss and the severity of the SLE were found not to be correlated. Therefore, the researchers established that

greater organ involvement does not necessarily mean that a greater degree of hearing loss will manifest.

Roverano et al. (2006) reported similar outcomes. The Systemic Lupus Erythematosus Disease Activity Index was administered prior to completing audiometric measures in order to quantify the disease activity of each patient. Of the 21 patients with a hearing loss, 11 reported little disease activity while the remaining 10 described persistent activity, experiencing a flare-up, or impairment caused by the disease activity. Roverano et al. (2006) concluded that there was not a correlation between disease activity and the presence of a hearing loss.

Numerous characteristics are associated with SNHL loss due to SLE. While there is variability in the outcomes, audiologists can anticipate a number of features yet still be vigilant of the possible dissimilarities. Bilateral, symmetrical, high-frequency sensorineural losses are likely to be the most common audiometric configuration. However, the audiologist should be aware of disease involvement if a loss in the mid or lower frequencies has been identified. Prior to the evaluation, the audiologist should ask the patient when the first symptoms of the disease were apparent. This is a useful additional piece of information that can be used by the audiologist. This interpretation of the published work suggests that there is a common model of hearing loss associated with SLE; however, audiologists should also be conscientious of the diverse audiometric features that can occur.

### **Vertigo**

Over the years, the connection between SLE and vertigo has become more apparent. Karatas et al. (2007), with the use of electronystagmography (ENG), found that out of the 19 patients reporting audiovestibular symptoms, eight of this subset

experienced symptoms of vertigo. The ENG testing established that 50% of the SLE patients had abnormal vestibular responses. Further evaluation identified the following abnormalities: gaze nystagmus, abnormal optokinetic performance, and positional nystagmus. Eleven of these 19 patients had poor caloric responses.

During this same period of time, Gomides et al. (2007), with the use of the Systemic Lupus Erythematosus Disease Activity Index, identified that 31.1% of their participants reported vertigo. This finding is significantly greater than the reports of vertigo from the control group (6.7%). Batuecas-Caletrío et al. (2013) recruited 89 SLE patients and determined that a subset of this population is susceptible to vestibular disorders. Peripheral vertigo was identified in eight patients and benign paroxysmal positional vertigo in one patient.

Kariya et al. (2016) evaluated the pathogenesis of vertigo associate with SLE. Fifteen temporal bones from patients with SLE were used to examine the integrity of the hair cells within the vestibular system, specifically in the saccular macula, the utricular macula, as well as within the cristae. When compared to the control group, there was a significant decrease in the number of type I hair cells, afferent cells, in all of the vestibular structures mentioned. However, there was not a significant difference when type II hair cells, or efferent cells, were evaluated.

### **Tinnitus**

The SLE patients have also reported the presence of tinnitus. When surveys were administered to participants, Maciazczyk et al. (2011) determined that 40% reported tinnitus. Furthermore, Karatas et al. (2007) identified nine individuals with tinnitus out of the 19 patients reporting audiovestibular involvement. While the reports

of tinnitus vary by patient, these studies indicate an association between tinnitus and SLE.

### **Identification of Autoimmune-Related Hearing Loss**

Standardized conventional pure-tone audiometry has a role in identifying the degree and type of hearing loss in SLE patients; however, additional diagnostic measures can be utilized to identify and monitor the progression of the hearing loss. This includes the administration of OAEs and extended high-frequency audiometry (EHFA). The following section will review the components of the standard audiometric battery as well as OAEs and high-frequency audiometry; all of which will aid the audiologist in the identification of secondary autoimmune inner ear disease due to SLE.

### **Audiometric Test Battery**

Otoscopy, immittance audiometry, pure-tone audiometry, and speech recognition testing comprise the collection of tests that are administered within a standard hearing evaluation. The purpose of otoscopy is to evaluate the condition of the outer ear and identify foreign bodies or pathologies. Immittance audiometry evaluates the integrity of the middle ear structures and measures the efficacy of how sound is transmitted within the outer and middle ear regions. Together, otoscopy and immittance audiometry provide early indications of possible conductive components.

Pure-tone audiometry is the subjective measurement of the transmission of tones from the outer ear to the cortical pathways leading to the auditory cortex. Auditory thresholds from 250 Hz to 8000 Hz are determined with the use of air-conduction testing, using insert or supra-aural headphones, and bone-conduction testing, with the utilization of a bone oscillator placed on the mastoid or forehead. If

thresholds are found outside of normal limits in an adult, exceeding a 20 dB hearing level, and conductive components are not apparent, this is evident of a pathology or condition within the inner ear or the central nervous system. Electrophysiological testing, such as ABR and electrocochleography, is completed to identify the functionality of these anatomical regions but is not part of the standard audiometric test battery.

### **Use of Otoacoustic Emissions**

Numerous researchers have identified autoimmune-related hearing loss with the utilization of transient evoked and distortion production OAEs. The purpose of OAE testing is to evaluate the functionality and motility of the cochlear outer hair cells (OHC). The anatomy of the organ of Corti permits the OHCs to move in a fashion that results in additional movement of nearby anatomical structures, such as at the basilar membrane, and is referred to as the electromotile response. This response that is elicited from an external sound source causes a wave of energy to travel in a distal direction from the cochlea and towards the external auditory meatus. Due to the tonotopic organization of the cochlea, frequency specific information can be obtained by using a stimulus tone located near the target frequency on the basilar membrane (Glatke & Robinette, 2007).

The OAE testing is comprised of presenting a stimulus using a probe placed in the ear canal. During the presentation of the stimulus in distortion product otoacoustic emissions (DPOAE) testing, the miniscule cochlear response is measured simultaneously. Individuals with normal hearing sensitivity have strong responses indicating normal OHC motility; presence of the electrical potential of the stria vascularis, the structure that supplies nutrients to the OHCs, and the absence of any

conductive component that prohibits the transmission of auditory signals leading to and from the cochlea. Abnormal or absent responses may indicate damage to the OHCs, damage to the stria vascularis, or a conductive component such as impacted cerumen and otitis media (Glattke & Robinette, 2007). Two types of OAEs have been used to monitor autoimmune inner ear disease: TEOAEs and DPOAEs.

The frequencies that had decreased OAE responses varied according to the type of autoimmune disease. Despite this variance, high frequencies were found to be more susceptible. Karabulut et al. (2010) completed a study comparing 30 subjects without SLE to 26 patients with SLE. The researchers concluded that patients with SLE may have absent DPOAEs for frequencies at 750 and 6000 Hz and absent TEOAEs at 2000 and 3000 Hz.

Larsen et al. (2015) obtained DPOAEs on rats with cochlear damage due to induced autoimmune involvement. The researchers completed DPOAEs on rats with cochlear damage due to autoimmune involvement. Absent responses were identified from 2000 to 63000 Hz. Therefore, in a controlled setting, Larsen et al. demonstrated that high-frequency regions of the cochlea were more susceptible to auditory damage and impaired DPOAEs.

Use of OAEs may be able to provide audiologists with the initial identification of auditory damage attributable to an abnormal autoimmune response in the cochlea. Botelho et al. (2014) recommended that the use of distortion product OAEs would be advantageous for monitoring early progression of the hearing loss. This method of testing could be especially beneficial for audiologists who are providing care to a high case load of patients. The use of OAEs provides a swift measurement of cochlear damage in relation to the functionality of the OHCs.



### **Use of Extended High-Frequency Audiometry**

The advantage of determining thresholds at frequencies above the conventional pure-tone test frequencies (250 – 8000 Hz) includes early identification of a hearing loss, especially with patients susceptible to ototoxic medications or with a history of noise exposure (Valiente et al., 2016). The extended high frequencies include 9000 to 20000 Hz. Karabulut et al. (2010) published their findings after completing thorough audiometric testing with 26 SLE patients matched with 30 healthy participants. The objective of this study was to fill in the gap in the literature pertaining to the knowledge of the mechanisms causing hearing loss in patients with SLE. Measures included EHFA (250, 500, 1000, 2000, 4000, 6000, 8000, 10000, 12000, 14000, and 16000 Hz), which was accompanied by DPOAE and TEOAE testing. When the SLE group and control group were compared according to pure-tone audiometry testing, significant differences between the SLE and control groups were identified at 250, 500, 1000, 2000, 10000, and 12000 Hz ( $p < 0.05$  significance). These findings indicate that a SNHL can extend into the higher frequencies, specifically to 10000 and 12000 Hz.

Lasso de la Vega et al. (2017) measured pure-tone thresholds from 125 to 8000 Hz, followed by EHFA pure-tone audiometry (9000, 10000, 11200, 12500, 14000, 16000, and 18000 Hz). Presbycusis was controlled for by recruiting younger patients (the mean age of the 55 participants was 41.5 years). Hearing thresholds obtained at 125-8000 Hz were compared to the EHFA that assessed 8000 to 18000 Hz. Findings revealed the presence of a SNHL between 8000 Hz and 18000 Hz in 70% of the patients. Significant threshold differences were identified at 10000, 11200, 12500,

16000, and 18000 Hz when the SLE and healthy participants were compared. Overall, it was determined that an extended high-frequency loss from 8,000 Hz to 18000 Hz was 33.6 times greater to be identified than a hearing loss that can be identified with traditional pure-tone audiometry testing. Lasso de la Vega et al. concluded their results with the following statement:

These results suggest and recommend that audiological assessment should be done not only with [pure tone average] but also [extended high-frequency audiometry] in patients with SLE in order to diagnose a possible subclinical hearing loss and modify the ongoing treatment or add a therapy to prevent a possible progression of hearing loss. (p. 165)

Hearing loss associated with autoimmune inner ear disease hearing loss should be monitored for a number of reasons. The first is to monitor the progression of the hearing loss over time as disease duration increases and in collaboration with physicians, select appropriate therapeutic options. Moreover, SLE patients should be monitored due to the risk of ototoxic effects of the treatment medications, such as the antimalarial drug hydroxychloroquine. The use of EHFA and OAEs allows the audiologist to document sub-clinical auditory damage as well as identify the early onset of the SNHL.

A suggested audiometric test battery for SLE patients needs to include the standardized pure-tone air and bone audiometry, impedance audiometry, speech and word recognition tests, as well as OAEs and EHFA. The addition of the OAEs and EHFA at the initial evaluation would establish baseline thresholds that could be used as a reference during the monitoring process. These measures may provide early warning signs of threshold changes related to the autoimmune disease.

### Ototoxic Medications

Multiple medications are prescribed to patients with SLE, and medications may be added depending on the secondary symptoms that accompany the disease. Synthetic antimalarial medications such as hydroxychloroquine (Plaquenil) and chloroquine (Aralen) are the primary drugs used to control the symptoms of SLE. Patients are counseled to take the medication for the remainder of their lives. These medications are also utilized in the treatment of rheumatoid arthritis and other connective tissue disorders (Bortoli & Santiago, 2007). What is concerning is the chemical relationship of these medications to quinine; both hydroxychloroquine and chloroquine are subtypes of quinine (Rynes & Park, 1993). Exposure to quinine, also used in the treatment of malaria, has been reported in patients diagnosed with SNHL, vertigo, and tinnitus (Jung et al., 1993). The ototoxic effects of quinine have been observed at the spiral ganglion neurons within the cochlea may also affect the mobility of the outer hair cells resulting in a decreased electromotile response (Zheng et al., 2001). At this time, toxic effects of quinine on the vestibular system have not been substantiated.

One case of accidental overdose of hydroxychloroquine resulted in permanent auditory and vestibular symptoms. Chansky and Werth (2017) reported an incident in which a patient experienced bilateral tinnitus and ataxia as well as symptoms of numbness in her arms and legs. Changes in hearing sensitivity were not noted.

The outcomes of several studies suggest that patients taking hydroxychloroquine and chloroquine can develop a sensorineural hearing loss. The earliest reported study was by Johansen and Gran (1998) who described the development of a severe SNHL loss in two SLE patients who had undergone years of

hydroxychloroquine drug therapy. Seckin et al. (2000) reported findings in which a woman developed a SNHL after five months of hydroxychloroquine treatment for rheumatoid arthritis. When hydroxychloroquine was prescribed as a drug therapy for a patient with HIV, a moderate-severe sensorineural hearing loss developed (Khalili et al., 2014). What is unique for this specific case is that when hydroxychloroquine exposure was ceased, audiometric testing revealed improved thresholds and indicated that for HIV patients, hearing loss may be reversible. While not commonly reported, physicians should be aware of the potential ototoxicity of the prescribed medication, especially due to the long-term treatment course.

The SLE patients are susceptible to SNHL due to progression of autoimmune inner ear disease, as well as the potential ototoxicity from antimalarial medications used to control the progression of SLE. This review suggests that audiologists should complete a thorough case history of SLE patients, identifying whether patients have been prescribed this class of medications and if so, the duration of the exposure. With the possible association of hydroxychloroquine and chloroquine to ototoxicity, audiologists should implement auditory and vestibular evaluations to monitor any changes in hearing sensitivity or balance

### **Resources and Support Groups**

The invasive nature of SLE in addition to multiple secondary disorders can be tolling on the patient. Support groups for SLE patients are available in multiple formats and offered across the United States. The Lupus Foundation of America has 17 chapters located in numerous states. Another national group is Molly's Fund Fighting Lupus which has chapters established in Oregon, Wyoming, Tennessee, and Massachusetts. Innumerable support groups are accessible online as well via websites

or Facebook. Lupus Warriors, a Facebook group, currently has 11,000 members and serves as a location to ask lupus-related questions and ask for support throughout difficult times. The website group Daily Strength currently has 874 members. The individuals are comprised of SLE patients and family members of those with SLE.

Partnerships have been established to advance lupus research, such as the Lupus Research Alliance. Located in New York City, the Lupus Research Institute promotes opportunities for SLE patients to participate in ongoing research with a focus on preventing, treating, and curing SLE. In June 2017, the Lupus Research Alliance established an advocacy group that successfully supported the expansion of funds for medical research on SLE. The opportunity to support other SLE patients and participants in ongoing research has formed a community for both patients and family members impacted by the disease. These resources assist patients recently diagnosed with hearing loss and provide information about other patients with similar experiences.

### **Future Research**

Future research should be qualitative in design in order to gain insight about the experiences of practicing audiologists caring for SLE patients, as each case may be different due to the nature of the disease. Investigation should be completed in order to determine what resources audiologists rely on. Furthermore, with the rise of studies evaluating the details and relationship between hearing loss and SLE, there is the question of whether this information has been integrated into graduate school courses and classes developed for audiologists to earn continuing education units. The current study answered these questions in a qualitative manner.

## CHAPTER III

### METHODS

The purpose of this chapter is to outline the steps of survey administration and data collection. The survey was constructed to describe the knowledge, attitudes, and practices of audiologists and audiology graduate students caring for patients with a hearing loss induced by systemic lupus erythematosus (SLE). This qualitative survey was intended to accumulate statements and information regarding the provision of audiological care for SLE patients and further inform best clinical practice in the future. Appendix A includes the Institutional Review Board approval of the survey and the overall project from the University of Northern Colorado.

#### **Development of the Survey Instrument**

Four sections made up the survey: demographics, knowledge, attitudes, and practices. The demographic questions developed for the audiologists inquired on the period of time the participant had practiced in audiology, what setting in which the participant was currently practicing in, and which state in the United States they were currently located. For graduate students, demographic questions included the number of years completed within the program, the number of years of supervised clinical experience accumulated, whether they were currently working at an internship or externship location, and the location of the educational institution.

Prior to the administration of the electronic survey, a small pilot study was completed to ensure that the survey was functional and that the questions were clearly

understood. The necessary modifications to questions were made after the trial period and before electronic links were distributed to participants in the main study.

### **Participants**

Two populations were invited to complete the survey (see Appendix B): graduate students enrolled in an accredited doctor of audiology graduate program or a doctor of philosophy graduate program in audiology, and licensed audiologists who were currently practicing. Licensed audiologists were included who were employed within a medical setting, speech and hearing clinic, corporate audiology group practice, otolaryngologist office, government patient care facility (such as Veterans Affairs or Indian Health Services), hospital, military setting, or private practice setting. Additional inclusion criteria considered whether the practicing audiologist provided direct patient care for an adult population. Audiologists were excluded who did not have experience seeing patients, were retired, or who were working in the educational field. Audiology graduate students had to be enrolled in an institution within the United States and the practicing audiologists must have been licensed in the United States.

### **Survey Instrument**

The electronic survey was developed using the Qualtrics (Smith, 2005) software. A number of strengths of this software include permitting the survey developer to coordinate the order in which the questions were sequenced based upon the responses submitted by the survey-taker. For instance, the question, “What best describes your current role in audiology?” served to identify the two groups of participants; practicing audiologists and graduate students. In relation to the response provided, the survey used skip logic to guide students to specific questions relating to

their academic experience, while certified practicing audiologists were directed to questions regarding their clinical experience.

Fifty-three questions were developed and divided into four sections: Demographics, Knowledge of Systemic Lupus Erythematosus, Attitudes Towards Systemic Lupus Erythematosus, and Practices in the Audiological Care of Patients with Systemic Lupus Erythematosus. Within the demographics portion, questions were presented according to whether the participant identified as a graduate student or a practicing audiologist. This was determined with the first question: “What best describes your current role in the field of audiology?” One purpose of the demographics section was to identify survey-takers who did not have a role in audiology and practicing audiologists who had not worked in a medical setting; both groups were directed to the end of the survey. Within the demographics portion, there were five questions for graduate students to answer and eight, at most, for practicing audiologists.

Sixteen questions made up the Knowledge of Systemic Lupus Erythematosus section that both participant groups completed. A total of 13 questions were formatted as forced-choice; three of these questions included text-boxes for the survey-taker to elaborate on the response. Furthermore, two questions required that the participant to select all of the options that applied to the statement presented.

The Attitudes Towards Systemic Lupus Erythematosus segment required answers to nine questions. The section began with a true/false question regarding the importance of referrals to otolaryngologists for SLE patients. This was then followed by two questions regarding whether additional tests procedures should be included in the test battery and which evaluations, from a provided list, should be included. A total



of eight statements and questions were presented that required the survey-taker to use a sliding scale to report an answer. Seven of these questions utilized a scale of *strongly disagree* to *strongly agree* and one question that utilized a not confident to confident scale.

The final section, Practices Associated with Systemic Lupus Erythematosus was composed of a collection of questions regarding whether the participant had cared for a patient with SLE, the referral process and what other professionals were typically contacted, whether the clinic in which the participant currently practiced had a protocol for these patients or other autoimmune disease patients, and open-ended questions about the development of a protocol for working with SLE patients.

### **Sampling Methods**

Survey promotion to both audiologists and audiology graduate students was electronic and accessible through a number of resources. One method was to post a brief description and survey link on audiology-related Facebook groups. These groups included Audiology Antics and Anecdotes For All Hearing Professionals, SOUNDing Board Hearing Healthcare Professionals, Academy of Doctors of Audiology, and Student Academy of Doctors of Audiology. All of these Facebook pages have practicing audiologists and audiology graduate student followers.

The survey was also promoted in the E-Newsletter SAAy Anything that is published every month. This allowed the survey to be distributed directly to graduate students.

Furthermore, the use of snowball sampling was used. After the audiologist or the graduate student completed the survey, a final note encouraged the survey-taker to share the survey link to colleagues and peers in order to promote snowball sampling.

Qualtrics (Smith, 2005) was programmed to only accept one survey response from an Internet protocol address to partially control for multiple responses to the same survey.

### **Descriptive Analysis**

Qualtrics was utilized to collect and analyze the data. This software permits a researcher to design an original survey tool as well as the ability to summarize and collate the results collected from the survey. Question development can range in format, including multiple-choice selections, text-box options, and a gap analysis style. In order to determine the SLE knowledge of both graduate students and practicing audiologists, the average number of questions correctly answered according to each participant group was calculated. For the attitudes portion, the multiple-choice questions were formatted to allow respondents to rate their answer from strongly disagree to strongly agree. The scores were averaged within each participant group and compared descriptively.

The goal of this analysis was to allow an interpretation of the results for both closed-choice questions and open-ended questions. The intention was to allow both respondent groups to provide details of particular experiences in caring for these patients and to gauge the degree of confidence and knowledge in caring for this SLE patient population

## CHAPTER IV

### RESULTS

#### **Respondent Characteristics**

A total of 133 individuals responded to the survey; 32 respondents identified as licensed audiologists and 86 as graduate students. The remaining 12 responses were false starts in which the survey was started but the first question, asking if the respondent was a licensed audiologist or graduate student, was not answered. Initial survey-based exclusion criteria in the licensed audiologist section included both whether the respondent had provided direct patient care after receiving their degree and if the individual was currently practicing in the United States. In the end, two respondents were excluded for not having experience with direct patient care. One observation made when implementing this exclusion criteria was that when the graduate students who answered no to the question “Have you read or heard about the autoimmune disease systemic lupus erythematosus?” many of them ( $n = 19$ ) stopped the survey shortly afterwards, and they may not have realized the survey would still record their responses to other questions.

Therefore, another consideration for analysis included the extent to which the survey was submitted with full completion. This chapter is composed of the information collected from the surveys in which the respondents answered all of the questions: 47 questions for licensed audiologists and 43 questions for graduate

students. This resulted in a total of 24 licensed audiologist and 53 graduate student surveys ( $n = 77$ ) used for analysis.

### **Demographics of Graduate Students**

The majority of respondents ( $n = 53$ ) who identified as graduate students reported that they were between their third and fourth year of the audiology doctoral program (33.96%,  $n = 18$ ), followed by those in between the second and third year (24.53%,  $n = 13$ ). Two to three years of clinical experience was the greatest amount (5.85%,  $n = 19$ ) reported, followed by three to four years (24.53%,  $n = 13$ ). See Table 1 for additional training and experience outcomes. Furthermore, when asked whether the externship had been started, 52.82% ( $n = 28$ ) of the respondents had not started this phase of the graduate program, while 47.17% ( $n = 25$ ) reportedly were at an externship site. This is considering the fact that some graduate students may start their clinical experience at a different time compared to students enrolled in other graduate programs.

Students were primarily in programs located in Colorado, South Dakota, and Wisconsin. Other locations included North Carolina, Oregon, and Texas. Overall, the respondents were completing programs in the Midwestern United States.

**Table 1***Program Status of Graduate Student Respondents*

Status	<1 yr %	1-<2 yrs %	2-< yrs %	3-<4 yrs %	4-<5 yrs %	5-<6 yrs %
Yr in program (n = 53)	9.43 (n = 5)	16.98 (n = 9)	24.53 (n = 13)	33.96 (n = 18)	13.21 (n = 7)	1.89 (n = 1)
Number of clinical yrs (n = 53)	13.21 (n = 7)	15.09 (n = 8)	35.85 (n = 19)	24.53 (n = 13)	9.43 (n = 5)	1.89 (n = 1)

**Demographics of Licensed Audiologists**

When posed with the question of terminal degree, 20 individuals (83.33%) reported to have a doctoral degree in audiology. Two individuals identified as having a master's of science (8.33%). Other degrees noted were one doctor of philosophy (4.16%) and one doctor of education (4.16%).

The location of the licensed audiologists was predominately Colorado (33.33%, n = 8) followed by Oregon (12.5%, n = 3) and North Carolina (12.5%, n = 3). Other locations to note were Texas (8.33%, n = 2), Minnesota (8.33%, n = 2), and Alabama (4.17%, n = 1). Pennsylvania, Idaho, Utah, Kentucky, and Maryland were also included with one individual from each.

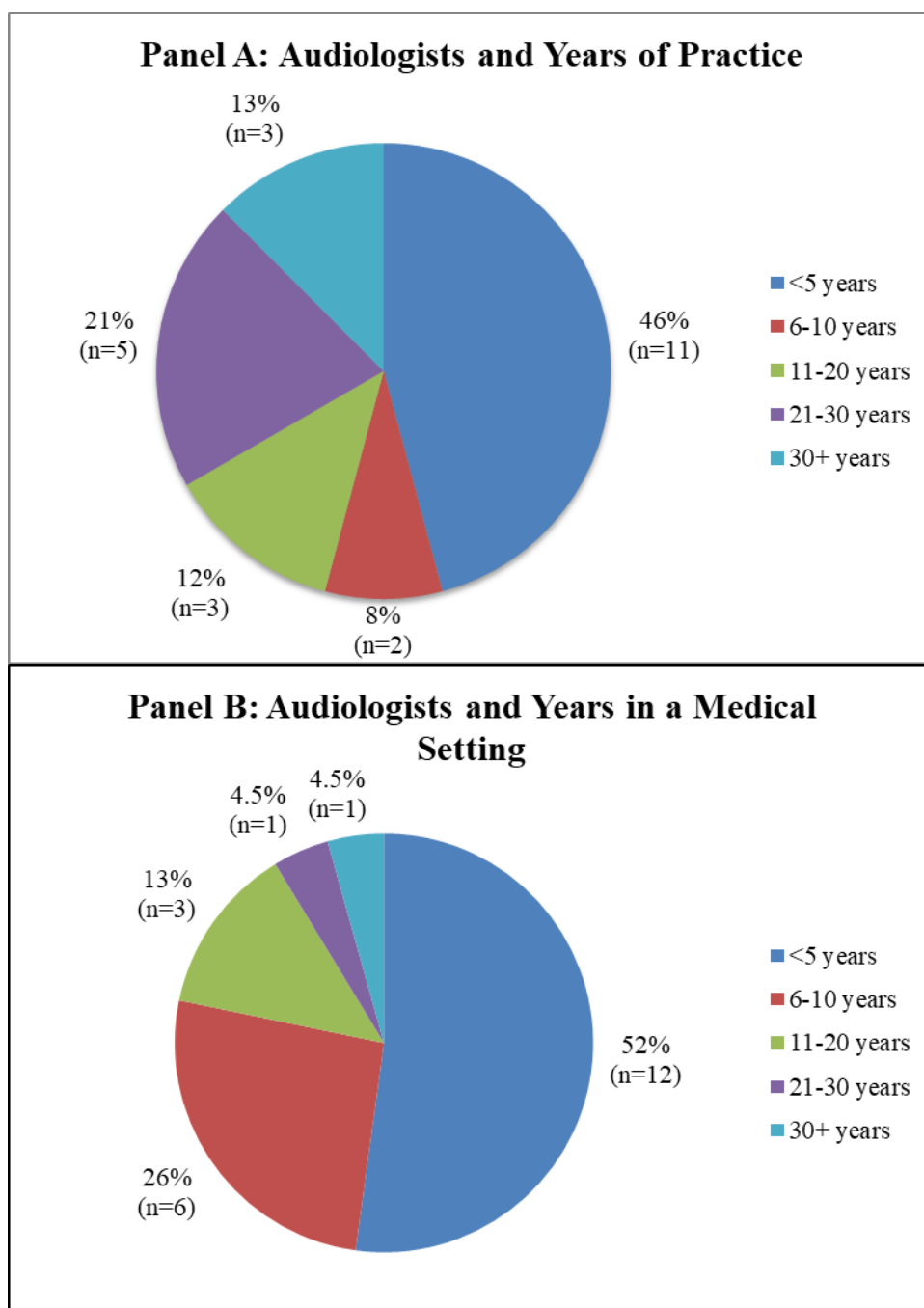
Licensed audiology respondents identified predominately as having practiced less than five years (46%, n = 11) followed by audiologists who have practiced for 21 to 30 years (13%, n = 5). The responses for the number of years in a medical setting

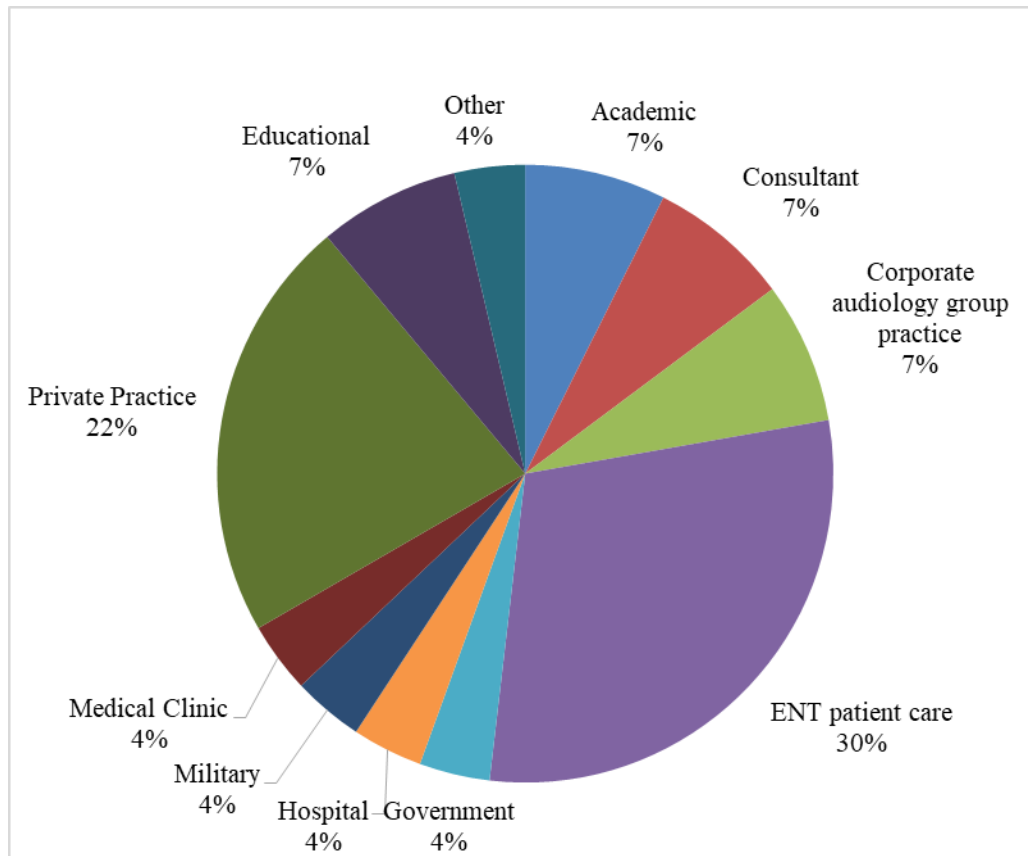
were mainly under five years (52%,  $n = 12$ ) and six to 10 years (26%,  $n = 6$ ). When asked whether the individual had practiced in a medical setting, most licensed audiologists had this experience when they took the survey (95.83%,  $n = 23$ ). All 24 respondents reported that they had provided some form of direct patient care after graduation. See Figure 1 for further details.

The most common current audiology practice settings included direct patient care in an ear, nose, and throat office (30%,  $n = 8$ ) and private practice (22%,  $n = 6$ ). This question was formatted for the respondent to select all the options that currently applied in the case that the respondent was working multiple part-time positions. One individual selected other and elaborated that they were employed at a nursing facility (4.17%) (see Figure 2).

**Figure 1**

*Number of Cumulative Years of Practice and Number of Years in a Medical Setting Reported by Audiologist Respondents*



**Figure 2***Practice Setting of the Licensed Audiologists***Knowledge Outcomes**

The remainder of this chapter is divided into three sections: knowledge, attitudes, and practices. Both audiologists and graduate student responses are separately integrated into the subsequent tables and figures. The knowledge section begins with questions regarding pre-existing knowledge of systemic lupus erythematosus (SLE) of both groups and followed by pre-existing knowledge of SLE and hearing loss. Outcomes of questions concentrating on clinical audiologic features of SLE and hearing loss are included at the end.



### Pre-existing Knowledge of Systemic Lupus Erythematosus

When posed with the question regarding whether students and audiologists were aware of SLE and SLE associated with hearing loss, more students reported awareness of SLE associated with hearing loss over SLE itself. In comparison, the 20 audiologists (83.00%) who identified as having an awareness of SLE were also aware of the diseases' association with hearing loss. The majority of both respondent groups had some degree of awareness regarding an association of SLE and hearing loss. A summary of the details are provided in Table 2.

**Table 2**

*Awareness of Systemic Lupus Erythematosus (SLE) and Awareness of SLE Related Hearing Loss by Graduate Students and Audiologists*

Knowledge of SLE	Graduate students ( <i>n</i> = 53) %	Licensed audiologists ( <i>n</i> = 24) %
Aware of SLE	60.38 ( <i>n</i> = 32)	83.00 ( <i>n</i> = 20)
Awareness of SLE and hearing loss	69.81 ( <i>n</i> = 37)	83.00 ( <i>n</i> = 20)

After establishing whether the respondents had this pre-existing knowledge, those who were knowledgeable of SLE were asked where they first learned of the disease. As a result, the pool of affirmative responses (*n* = 52) was smaller than the total number of respondents (*n* = 77). A total of 32 graduate students and 20 licensed

audiologists were included and responses were spread across all sources of information. Graduate students primarily had a personal relationship as their source of initial knowledge at 21.88% ( $n = 7$ ). Audiologists, on the other hand, initially learned of SLE from providing care to a patient with 15% ( $n = 3$ ) or from another health care professional at 15% ( $n = 3$ ). Graduate students also reported learning of SLE in high school with 3.13% ( $n = 1$ ), and one respondent reported having an autoimmune disorder related to SLE at 3.13% ( $n = 1$ ). For the licensed audiologists, one was diagnosed with SLE (5%), another needed SLE to be ruled out as their own personal medical condition (5%), and the third from their mother (5%); it was not noted whether the mother was diagnosed with SLE. A summary is provided in Table 3.

For knowledge of hearing loss related to SLE, this was again directed to the separated pool of 52 respondents. A total of 35.85% ( $n = 19$ ) graduate students reportedly learned of the association from a graduate school course. These courses included standard medical audiology classes, such as medical aspects, pathologies of hearing, and auditory and vestibular pathologies. One student (3.13%) reportedly had a pediatric audiology class that covered this topic while another student (3.13%) had a clinical problem-solving class that included SLE. A similar outcome was also evident with audiologists. Graduate school courses that were not the standard medical audiology class included a case study in a practicum staffing course (see Table 4).

**Table 3**

*Sources of Systemic Lupus Erythematosus (SLE) Information Identified by Graduate Students and Audiologists*

Source of information	Graduate students (n = 32) %	Audiologists (n = 20) %	Total responses (n = 52) %
Electronic media	6.25 (n = 2)	0.00 (n = 0)	3.85 (n = 2)
From an audiology colleague	6.25 (n = 2)	10.00 (n = 2)	7.69 (n = 4)
From another health care professional	9.38 (n = 3)	15.00 (n = 3)	11.54 (n = 6)
Graduate school course	18.75 (n = 6)	10.00 (n = 2)	15.39 (n = 8)
Peer reviewed journal	3.13 (n = 1)	0.00 (n = 0)	1.92 (n = 1)
Personal relationship	21.88 (n = 7)	0.25 (n = 5)	23.07 (n = 12)
Print, books, newspaper, magazine	3.13 (n = 1)	0.00 (n = 0)	1.92 (n = 1)
Providing care to a patient with SLE	3.13 (n = 1)	15.00 (n = 3)	7.69 (n = 4)
Television	18.75 (n = 6)	10.00 (n = 2)	15.39 (n = 8)
Other	9.38 (n = 3)	15.00 (n = 3)	11.54 (n = 6)

**Table 4**

*How Respondents were First Aware of the Relationship of Hearing Loss to Systemic Lupus Erythematosus (SLE)*

Responses	Graduate students (n = 53) %	Audiologists (n = 24) %	Total (n = 77) %
Audiologic care to patient with SLE	1.89 (n = 1)	16.67 (n = 4)	6.49 (n = 5)
Audiology colleague	11.32 (n = 6)	8.33 (n = 2)	10.39 (n = 8)
Electronic media/internet source	5.66 (n = 3)	4.17 (n = 1)	5.19 (n = 5)
Graduate school course	35.85 (n = 19)	20.83 (n = 5)	31.17 (n = 24)
Healthcare professional	3.77% (n=2)	4.17% (n=1)	3.90% (n=3)
Peer reviewed journal	1.89 (n = 1)	12.5 (n = 3)	5.19 (n = 4)
Personal relationship	1.89 (n = 1)	4.17 (n = 1)	2.60 (n = 2)
Print, books, newspapers, magazine	0.00 (n = 0)	0.00 (n = 0)	0.00 (n = 0)
Television	1.89 (n = 1)	4.17 (n = 1)	2.60 (n = 2)
Were not aware	30.19 (n = 16)	16.67 (n = 4)	25.97 (n = 20)
Other	5.66 (n = 3)	8.33 (n = 2)	6.49 (n = 5)

Figure 3 summarizes the sub-set of responses for when respondents first became aware of SLE. Graduate students were generally aware of SLE for less than five years as a predominant answer (63%,  $n = 20$ ) which coincides with their years in the degree program. For audiologists, 25% ( $n = 5$ ) reported that it had been six to 10 years since they had first become aware of SLE and 25% ( $n = 5$ ) selected that they were unsure of the time frame.

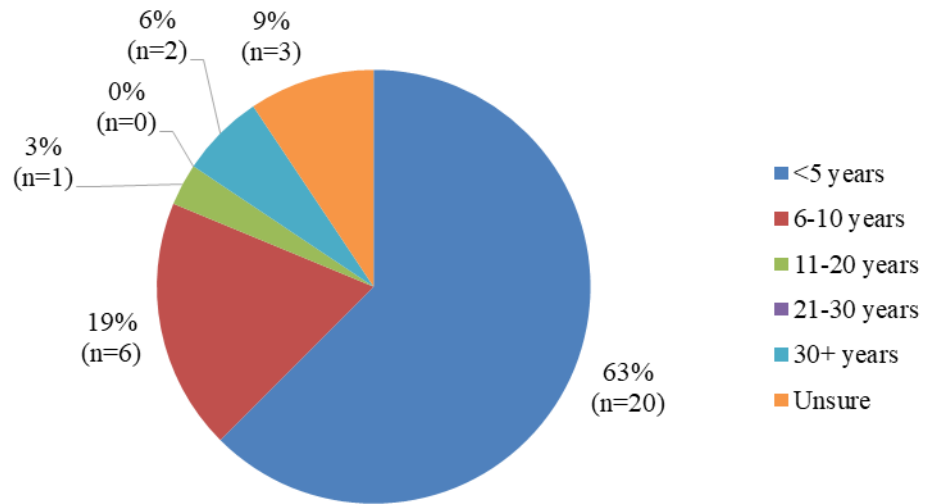
### **Knowledge of Audiologic Symptoms**

The following section outlines how graduate students and licensed audiologists were able to answer questions specifically regarding audiologic symptoms observed in patients with SLE. Due to a survey design error that permitted the respondent to skip the question, two questions were removed due to the high rate of skipped responses. These questions included asking what the greatest degree of hearing loss tends to be in patients with SLE and the question on whether the hearing loss tends to be progressive or stable. Eight knowledge questions remained afterwards; Table 5 provides a summary of these results in the context of correctness based on peer-reviewed evidence in the literature (Chapter II).

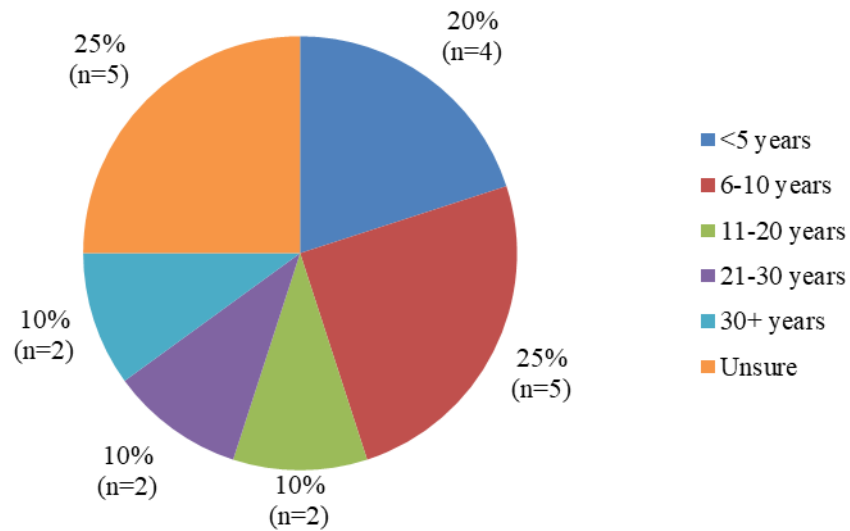
**Figure 3**

*How Long Ago were Graduate Students and Licensed Audiologists First Aware of Systemic Lupus Erythematosus (SLE)*

**Panel A: When Graduate Students Were First Aware of SLE**



**Panel B: When Audiologists Were First Aware of SLE**



**Table 5***Number of Correct Responses between Graduate Students and Licensed Audiologists*

Response	% correct	
	Graduate students (n = 46)	Licensed audiologists (n = 24)
What type of hearing loss is commonly associated with hearing loss from SLE autoimmune responses?	86.79 (n = 46)	87.5 (n = 21)
For patients with SLE, does the hearing loss tend to be bilateral or unilateral?	41.51 (n = 22)	37.5 (n = 9)
Do you believe that the duration of SLE and the degree of hearing loss are correlated? (e.g., the longer a person has had SLE, the poorer the hearing).	71.70 (n = 38)	62.5 (n = 15)
Can patients with SLE experience vertigo that is attributed to the presence of the autoimmune disease?	75.47 (n = 40)	70.83 (n = 17)
SLE is thought to arise in patients from which of the following factors:	77.36 (n = 41)	66.67 (n = 16)
The following medications are used in the treatment of SLE. Based on your knowledge, which ones can be ototoxic, if any? Selected choice	41.51 (n = 22)	37.5 (n = 9)

*Note.* SLE = systemic lupus erythematosus.

### **Attitudes of Licensed Audiologists and Audiology Graduate Students**

Both groups were presented the question on whether additional audiological tests should be added to the standard audiometric test battery. This is specific to the situation in which a patient with SLE is being provided audiologic care. The majority of both graduate students and audiologists (84.42%,  $n = 65$ ) held the opinion that additional tests should be included: 92.45% ( $n = 48$ ) with graduate students and 70.83% ( $n = 17$ ) with licensed audiologists.

Respondents were allowed to select multiple tests. Evaluation with an extended high frequency audiometry (EHFA) and distortion product otoacoustic emissions (DPOAEs), were common selections within both groups; however, a total of 28 graduate students (58.33%) selected EHFA more often than the eight audiologists (47.09%). See Table 6 for the number of total responses associated with each test.

Figure 4 compares graduate student and audiologist responses from the Likert scale questions regarding respondent attitudes. Neither graduate students nor audiologist feel confident in their knowledge of SLE or their ability to counsel patients with SLE. Audiologists and graduate students were neutral regarding whether they had the resources to allow them to provide audiologic care to patients with SLE.



**Table 6**

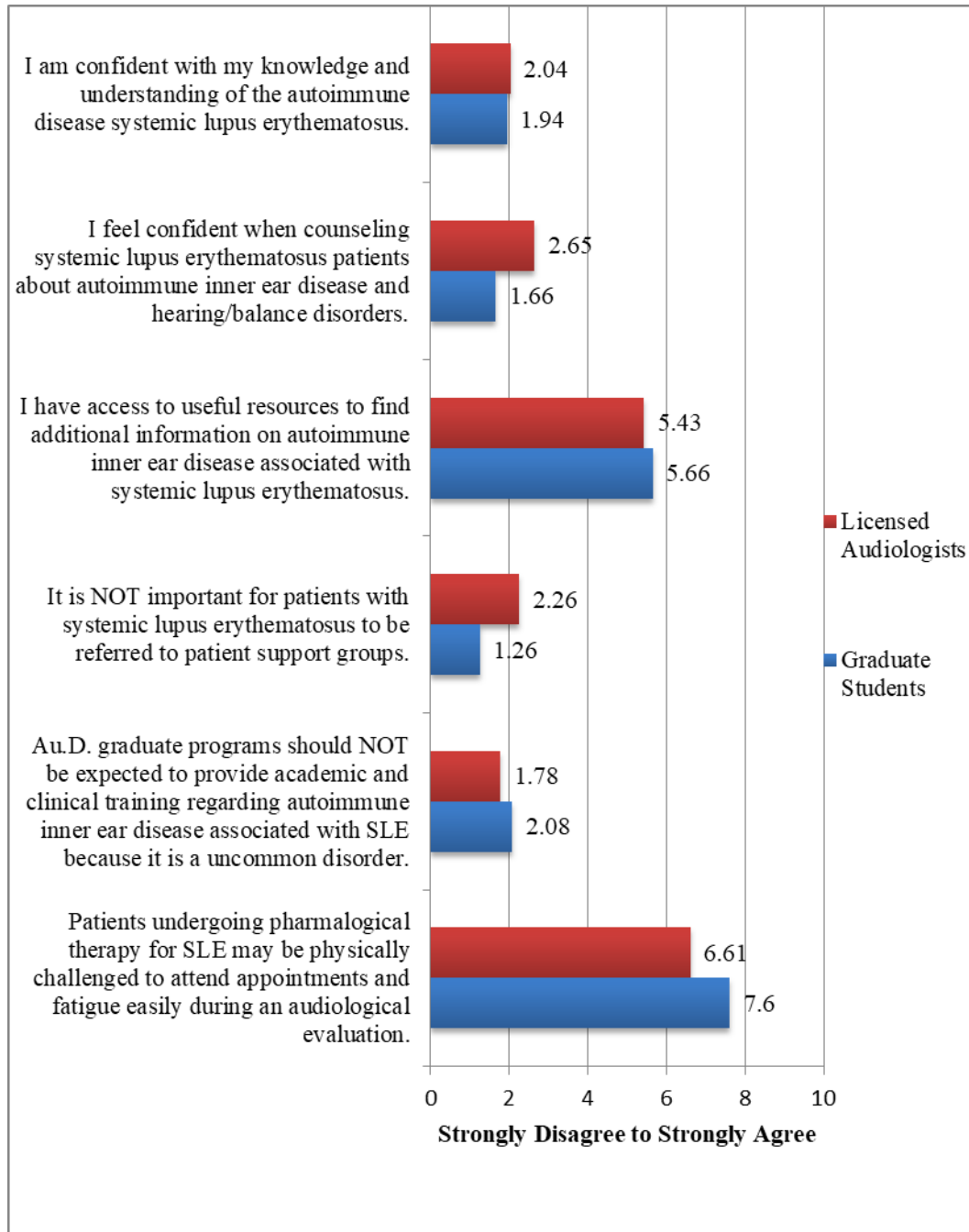
*Tests that Should be Added to the Standard Audiometric Battery for Patients Being Monitored for Systemic Lupus Erythematosus (SLE)*

Test	Graduate students (n = 48) %	Audiologists (n = 17) %	Total (n = 65) %
Acoustic reflex threshold	54.17 (n = 26)	58.82 (n = 10)	55.38 (n = 36)
Acoustic reflex decay	22.92 (n = 11)	23.53 (n = 4)	23.08 (n = 15)
APD Testing	2.08 (n = 1)	0.00 (n = 0)	1.54 (n = 1)
Click ABR	45.83 (n = 22)	29.41 (n = 5)	41.54 (n = 27)
DPOAEs	64.58 (n = 31)	82.35 (n = 14)	69.23 (n = 45)
ECohG	20.83 (n = 10)	17.65 (n = 3)	20.00 (n = 13)
Extended HFA	58.33 (n = 28)	47.09 (n = 8)	55.38 (n = 36)
Extended HF DPOAEs	39.58 (n = 19)	23.53 (n = 4)	35.38 (n = 23)
Immittance testing	25.00% (n = 12)	11.76% (n = 2)	21.54% (n = 14)
Multi-frequency tympanometry	2.08 (n = 1)	0.00 (n = 0)	1.54 (n = 1)
Speech in noise	0.00 (n = 0)	11.76 (n = 2)	3.08 (n = 2)
TEOAEs	12.50 (n = 6)	11.76 (n = 2)	12.31 (n = 8)
VEMP	2.08 (n = 1)	0.00 (n = 0)	1.54 (n = 1)
VNG	4.17 (n = 2)	0.00 (n = 0)	3.08 (n = 2)
Wide-band reflectance	4.17 (n = 2)	0.00 (n = 0)	3.08 (n = 2)

*Note.* APD = auditory processing disorder, ABR = auditory brainstem response, DPOAEs = distortion product otoacoustic emissions, ECohG = electrocochleography, HFA = high frequency audiometry, TEOAEs = transient evoked otoacoustic emissions, VEMP = vestibular evoked myogenic potential, VNG = videonystagmography.

**Figure 4**

*Comparison of Mean Likert Scores between Graduate Students and Licensed Audiologists.*



Both groups felt strongly that patients with SLE should be referred to additional support groups and that doctor of audiology graduate students should have access to materials on the disease and hearing loss. Both groups generally agreed that patients undergoing pharmacologic treatment may fatigue easily during testing.

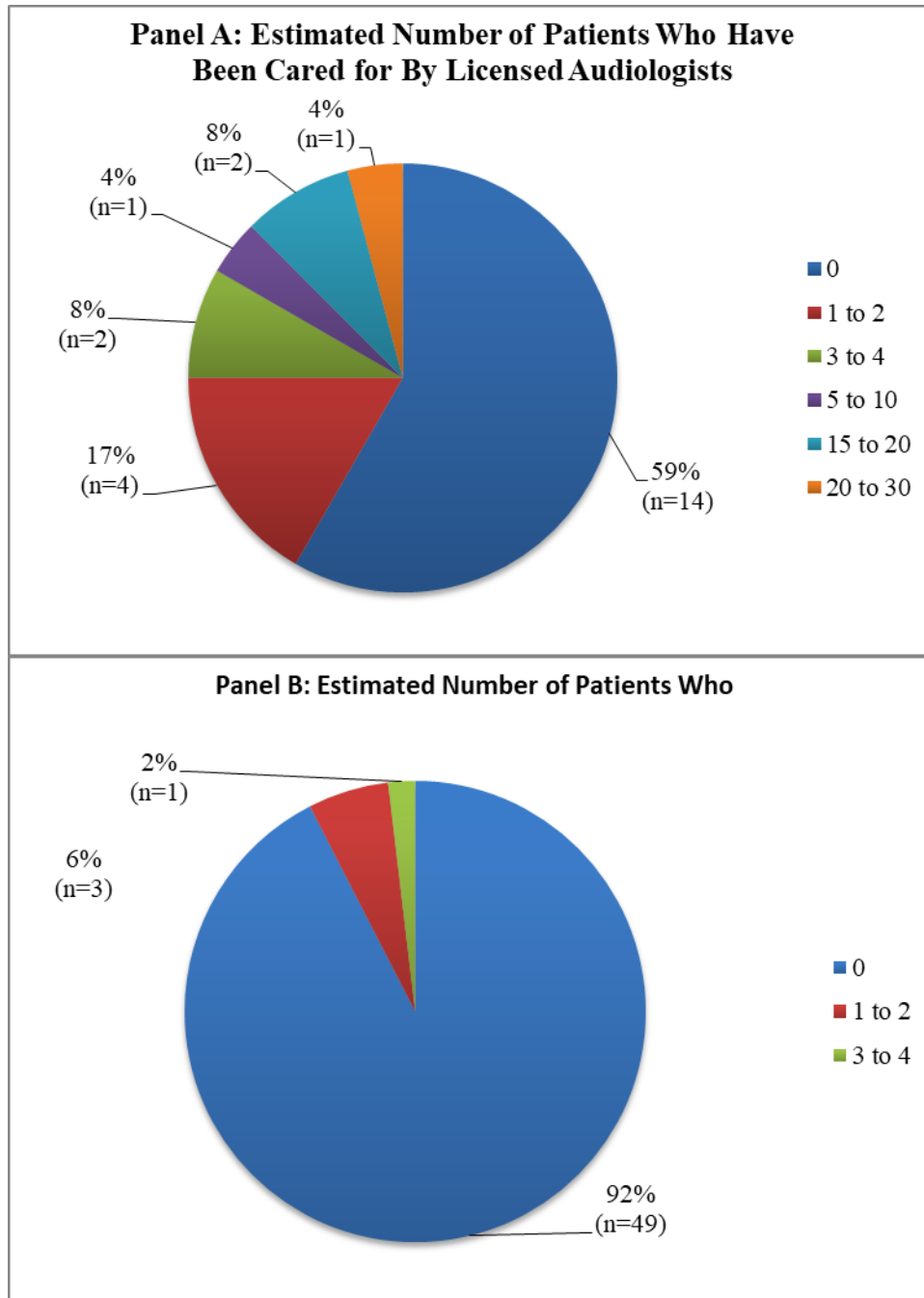
### **Clinical Practices for Systemic Lupus Erythematosus Patients**

The majority of both groups of respondents had not provided care to patients with SLE. However, 92% ( $n = 19$ ) of graduate students had no experience with SLE patients, compared to 59% ( $n = 14$ ) of audiologists. See Figure 5 for an in-depth representation of their experiences.

The sub-set of respondents ( $n = 14$ ) who had provided audiologic care to patients with SLE differed with regard to how often hearing evaluations for patients with SLE should be advised (see Table 7). Graduate students were equally distributed between annually, every 6 months, and every 3 months (25%,  $n = 1$ ). The audiologists (70%,  $n = 7$ ) were more likely to recommend annual evaluations. Twenty to 25% of respondents for both groups acknowledged that annual evaluations were sufficient unless changes in hearing were subjectively experienced by the patient.

**Figure 5**

*Estimated Number of Patients with Systemic Lupus Erythematosus (SLE) Who have been Cared for with Licensed Audiologists and Graduate Students*



**Table 7**

*Response Summary to the Survey Question “How Often Do You Recommend Patients with Systemic Lupus Erythematosus to Have a Hearing Evaluation?”*

Recommendation	Graduate students (n = 4) %	Audiologists (n = 10) %	Total (n = 14) %
Annually	25 (n = 1)	70 (n = 7)	57.14 (n = 8)
Every 6 months	25 (n = 1)	10 (n = 1)	14.29 (n = 2)
Every 3 months	25 (n = 1)	0 (n = 0)	7.14 (n = 1)
Every month	0 (n = 0)	0 (n = 0)	0.00 (n = 0)
Annually, sooner if changes perceived	25 (n = 1)	20 (n = 2)	21.43 (n = 3)

Table 8 summarizes the additional tests that are given to patients with SLE. A total of eight respondents contributed to this portion of the survey. It should be noted that these audiometric tests are not widely used within the field of audiology.

**Table 8**

*Additional Audiometric Rests Provided to Patients with Systemic Lupus Erythematosus?*

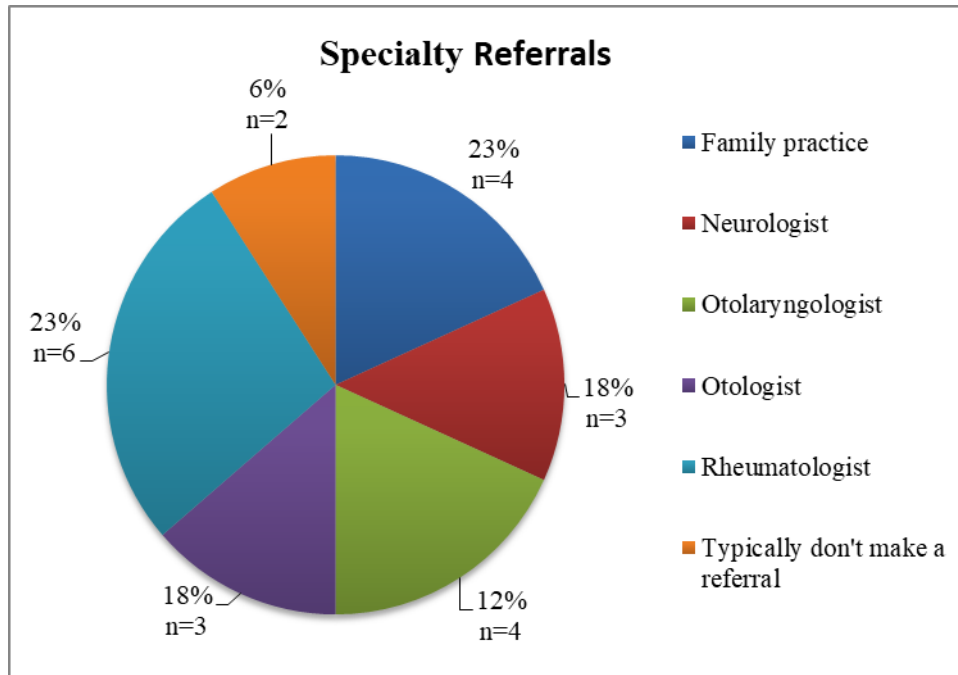
Recommendation	Graduate students ( <i>n</i> = 2)	Audiologists ( <i>n</i> = 6)	Total ( <i>n</i> = 8)
APD testing	( <i>n</i> = 1)	( <i>n</i> = 0)	( <i>n</i> = 1)
OAEs	( <i>n</i> = 0)	( <i>n</i> = 1)	( <i>n</i> = 1)
Speech in noise	( <i>n</i> = 0)	( <i>n</i> = 3)	( <i>n</i> = 3)
Tone decay	( <i>n</i> = 1)	( <i>n</i> = 0)	( <i>n</i> = 1)
Other			
My basic battery is PTA/PTB, speech thresholds/word recognition, DPOAEs, immittance...reflexes	( <i>n</i> = 0)	( <i>n</i> = 1)	( <i>n</i> = 1)
Decay, OAEs, ABR	( <i>n</i> = 0)	( <i>n</i> = 1)	( <i>n</i> = 1)

*Note.* ABR = auditory brainstem response, APD = auditory processing disorder, DPOAEs = distortion product otoacoustic emissions, OAEs = otoacoustic emissions, PTA = pure tone average, PTB = pure tone baseline.

Patient referrals made by licensed audiologists were predominately going to rheumatologists (23%, *n* = 6), (23%, *n* = 4) to family practice physicians, and (12%, *n* = 4) to otolaryngologist. This was followed by an otologist referral (18%, *n* = 3) and neurology with (18%, *n* = 3). Two graduate students had experience with referrals; one referred a patient with SLE to neurology, while the other student reported making a referral to rheumatology (see Figure 6).

**Figure 6**

*Summary of Referrals to Other Specialists Made by Audiology Graduate Students and Audiologists*



For the respondents who stated that a referral was not made, a follow up question requested further detail for their response. The licensed audiologist stated that there was already a team of providers in place and so no referral was necessary. This response was also reported by one of the two graduate students. The second graduate student had typed in an incoherent answer into the text box.

Two graduate students and two licensed audiologists stated that they use a specific clinical protocol for patients with SLE; however, no contact information was offered in order to learn more about the protocols (this was a voluntary question). There appears to be a strong professional interest of graduate students (89.58%,  $n = 43$ ) and licensed audiologists (95.00%,  $n = 19$ ) of having a clinical protocol

implemented for patient with SLE. This was also reflected in the question that allowed the respondents to select all of the resources they believed would be beneficial to have in the clinic followed by an SLE resource list (83.82%,  $n = 57$ ). See Table 9 for further details. Additionally, eight respondents (11.76%), four from the graduate student group and four audiologists, reported having additional training or continuing education in SLE.

**Table 9**

*Summary of Types of Information that Would be Useful to Graduate Students and Audiologists with Regard to Providing Audiologic Care to Patients with Systemic Lupus Erythematosus (SLE)*

Response	Graduate students ( $n = 48$ ) %	Audiologists ( $n = 20$ ) %	Total ( $n = 68$ ) %
Clinical diagnostic and monitoring protocol	89.58 ( $n = 43$ )	95.00 ( $n = 19$ )	91.18 ( $n = 62$ )
List of interdisciplinary team members	81.25 ( $n = 39$ )	50.00 ( $n = 10$ )	72.06 ( $n = 49$ )
SLE resource list for the clinician	83.33% ( $n = 43$ )	70% ( $n = 14$ )	83.82% ( $n = 57$ )
Support group contacts	85.42 ( $n = 41$ )	55 ( $n = 11$ )	76.47 ( $n = 52$ )
Other			
Contact info of specialists	0.00 ( $n = 0$ )	5.00 ( $n = 1$ )	5.00 ( $n = 1$ )
Signs and symptoms	0.00% ( $n = 0$ )	5.00% ( $n = 1$ )	5.00% ( $n = 1$ )



### Open-Ended Comments

The final question of the survey permitted the respondent to add questions or comments regarding the content of the survey. The comments are transcribed below:

“Is this the same as plain old lupus” (graduate student)?

We do not see many of these patients . . . but when we do it is important to know what evaluations to provide and be able to counsel these patients on the disorder and its progression. Additionally, it is essential to begin rehabilitative treatment ASAP [as soon as possible] and be able to explain why/how and what will be necessary in the future. (licensed audiologist)

“Most rheumatologists do not know or inform their patients that Plaquenil is ototoxic and worse for some patients than others and requires education and monitoring” (licensed audiologist).

Is systemic lupus erythematosus a specific type of lupus, or is it the medical name for lupus? I had to Google this. You could have put a brief explanation of what SLE [systemic lupus erythematosus] is at the start of your survey to clarify this point, but you didn't. Also, you could have had more questions with ‘I don't know’ as possible answers, because without that option I had to randomly choose something and I don't know how those guesses will bias your statistical analysis. (licensed audiologist)

Not a comment about providing care to patients, but rather feedback on the survey—You did not provide enough opportunity to respond “I don't know.” That may skew your results. When not provided the opportunity to respond “I don't know” I guessed. If the purpose of the study is to determine the state of knowledge of currently practicing audiologists, without “I don't know” as an option, you may overestimate the awareness of this condition. It may also be helpful for you to know whether someone practicing in a “medical setting” is practicing in an adult focused facility, a pediatric focused facility or a facility that provides services across the lifespan. Pediatric audiologists may not encounter this condition. (licensed audiologist)

### Summary

These outcomes have identified subject areas in which the respondents were knowledgeable about SLE and areas where improvement can occur. The general consensus is that audiologists and graduate students were aware of the disease and the

potential that a hearing loss of an autoimmune basis can develop. However, when it came to the concept of what additional services and monitoring this patient group may require, respondents were unsure and hesitant of their approach. For instance, respondents on average were more likely to disagree that they are confident in their understanding of SLE. This was also seen in with the counseling aspect, that the respondents were likely to disagree that they would be able to appropriately counsel someone with SLE.

When the respondents were permitted to select what audiometric tests to add to the standard test battery when a patient with SLE is seen for a hearing evaluation, EHFA and DPOE were the tests that were selected the most. Another takeaway from the responses is that a protocol is needed. When the provided the option, both groups responded that it would be useful in the clinic. Other resources were also requested, such as a resource list and a list of support groups, but the highest selected option was to have a universal protocol available. Overall, according to the survey responses there is an awareness of the disease but in terms of follow up, graduate students and licensed audiologists use a variety of methods to provide care to this patient population.

## CHAPTER V

### DISCUSSION

#### **Experience of Providing Care**

One component of this research was to identify the knowledge of hearing loss associated with systemic lupus erythematosus (SLE) within the groups of practicing audiologists and audiology graduate students. Respondents of both groups were generally aware of SLE and the relationship of SLE with hearing loss. Graduate students were more likely to report having first learned of SLE from a personal relationship. Practicing audiologists on the other hand reported learning of SLE from another health care professional.

Only 31% of both groups combined reported that they first learned of hearing loss associated with SLE from graduate school courses. Another factor to consider is that this topic area is fairly new in the field of audiology, bearing in mind that the majority of the published research on this topic has been within the past decade. For instance, the most recent prevalence studies that were completed by the Centers for Disease Control and Prevention were published between 2014 and 2017 (Dall'Era et al., 2017; Izmirly et al., 2017; Lim et al., 2014; Somers et al., 2014).

Overall, this is representative that some audiologists and graduate students may have to rely on first-hand clinical experiences or other healthcare providers to be introduced to this topic. The respondents also reported a lack of confidence in counseling patients with SLE. This may also relate back to the lack of formal training

in the disorder. Audiology graduate programs may need to expand their course content to include SLE as a relevant clinical pathology and equip audiologists to counsel SLE patients.

As indicated by this current research study, 59% of licensed audiologists and 92% of graduate students do not have experience providing care to patients with SLE. This is anticipated with graduate students, considering that these individuals are in the early stages of their clinical experience. Approximately 40% of licensed audiologists, on the other hand, did recall having this background. While it is not anticipated that the audiologist will be presented with a specific case such as this on a regular basis, the concern is that this may result in the clinician not taking necessary means of care specific to this population because of the irregular exposure. Considering that there is a moderate to high prevalence of hearing loss in patients with SLE, it is critical that the audiologist have resources to refer to if the time comes around that the audiologist sees a patient within this population (Rahne et al., 2017).

It was somewhat surprising in this digital era to note that electronic media, peer reviewed journal articles, or other print resources such as books were not more commonly flagged (~1%,  $n = 5$ ) as an initial source on SLE. It was beyond the scope of this research study to review available resources, but it may be useful to provide links to existing resources on professional organization websites and publicize on social media frequented by audiologists and audiology graduate students. An abbreviated list of resources is available in Appendix C that an audiologist can utilize.

Seven questions regarding expected audiologic symptoms and hearing loss characteristics were presented to determine the familiarity of graduate students and licensed audiologists with this subject area. These questions were forced-choice and

did not provide an “I don’t know” option. The purpose of this formatting was to capture what direction the respondent was leaning towards. An answer key to these questions has been included in Appendix D.

The majority of both groups were able to correctly identify that the hearing loss tends to be sensory or neural; 86% with graduate students and 87% of licensed audiologists that is displayed in Table 5. Similar outcomes were observed to the questions focusing on disease duration and degree of hearing loss, that vertigo can be a symptom, and whether SLE influence by genetic and environmental factors.

When it comes to these topic areas, audiologists and graduate students generally have the right mindset; however, this does not directly reflect how confident the respondents were in their selection. Reflecting on the outcomes of the sidler scales of the attitudes section, both groups believed that they did not have a high degree of confidence with their knowledge and understanding of SLE as well as the confidence in their skills to counsel a patient with SLE. As a consequence, the respondents were able to perform well with a subset of these questions; however, their confidence may be wavering.

The question on whether the hearing loss tends to be bilateral or unilateral reflected that respondents tended to select the bilateral only option: 41% of graduate students and 37% of licensed audiologists. Similar outcomes were associated with the questions on the susceptibility of patients with SLE to develop a hearing loss and correctly determining that hydroxychloroquine can be ototoxic. It appears that current audiologic resources may need to provide greater focus on these topic areas.

### **Need for Clinical Resources**

There was an overwhelming consensus from both respondent groups that additional SLE-related resources are needed in the clinic. There was an agreement from both groups that they were not confident with their knowledge and understanding of SLE.

### **Development of a Clinical Protocol**

Graduate students and practicing audiologists agreed that a clinical diagnostic and monitoring protocol would be desirable to guide the evaluation and care of patients with SLE. At this time, a protocol or guideline addressing the audiologic care of patients with SLE has not been developed by any professional organization (e.g., American Academy of Audiology or American Speech-Language Hearing Association). Professional assessment and monitoring guidelines would result in a peer-reviewed agreement of how this patient population should be cared for by audiologists. The existence of a peer-reviewed clinical protocol or guideline would provide an additional opportunity to educate audiologists and graduate students regarding hearing loss in patients with SLE. The following sections address the clinical components that might be integrated into the guideline.

### ***Case History***

The development of a protocol would not be absent of its challenges. Each patient with SLE can vary in system-wide symptoms and variability is expected with regard to auditory symptoms as well (Al-Sukaini et al., 2014). Furthermore, symptoms can change over time, requiring that regular comprehensive updates on patient symptoms to be updated at each visit.

The case history portion of the protocol should be as comprehensive as possible in the instance that symptoms change in the future. Questions should inquire about the time of SLE diagnosis, considering that patients diagnosed with SLE early in their lives exhibit significantly decreased auditory thresholds that are unrelated to presbycusis (Maciazczyk et al., 2011).

Moreover, features of tinnitus should be recorded at this time, as this could determine whether additional evaluations are needed. The research suggests that tinnitus symptoms are prevalent in patients with SLE; therefore, it should be documented as soon as possible (Karatat et al., 2007; Maciazczyk et al., 2011). With research suggesting nearly half of patients with SLE experiencing bilateral tinnitus, the audiologist should anticipate counseling the patient that the cause of the symptom is related to their autoimmune disease (Maciazczyk et al., 2011).

Vertigo is also a common feature. The audiologist needs to anticipate counseling the patient that the pathogenesis of the vertigo symptoms may be related to their SLE. The temporal bone study completed by Kariya et al. (2016) have supported this concept of disease vestibulotoxicity and Gomides et al. (2007) identified a rate of 31.1% of patients with SLE who were experiencing vestibular symptoms. As an audiologist, the case history should incorporate questions addressing any current vertigo features are experienced by the patient with SLE.

A case history question devoted to the identification of current medications should also be included. This could come in the simple form of a checklist to allow the audiologist to quickly review with the patient. A concerning finding from this present research is that respondents of both groups were not aware of the ototoxic effect of Plaquenil (Seckin et al., 2000). Incorporating a standard sub-set of questions related to

Plaquenil administration to the protocol would allow the audiologist to keep in mind of not only the immune factors at large, but also the potential need for ototoxicity monitoring.

Respondents expressed the importance of knowing the interdisciplinary team that is overseeing the patient. This is an additional section to be highlighted in the case history intake. What is paramount is determining the rheumatologist treating the patient, especially if the audiologist has concerns that need to be communicated to the physician related to changes in auditory functioning, especially in the context of ototoxicity.

***Additions to the Standard  
Audiometric Battery***

An SLE clinical protocol not only should provide guidelines to case history intake, an outline of the audiometric battery should also be included. The predominant view from respondents in terms of evaluating patients with SLE supported the inclusion of additional tests beyond the standard audiological test battery. A review of the literature suggests that an SLE protocol should include the additional test measurements of extended high-frequency audiometry (EHFA) and distortion product otoacoustic emissions (DPOAEs) (Botelho et al., 2014; Lasso de la Vega et al., 2017). Both of these tests permit the audiologist to identify early features of a sensorineural hearing loss and document any sub-clinical auditory damage. Furthermore, considering that the malarial pharmaceutical hydroxychloroquine or Plaquenil used with patients with SLE has the potential of causing the hearing loss, especially in the higher frequencies, these tests would offer the opportunity to monitor the patient in terms of ototoxicity (Chansky & Werth, 2017; Seckin et al., 2000).



Incorporating EHFA and DPOAEs would provide general information regarding neural functioning and an additional diagnostic measure to use if changes in hearing sensitivity occur. Published research has indicated that the hearing loss can become worse over time (Cordeschi et al., 2004; Sperling et al., 1996). Reflecting on the current published research, it is critical that clinicians within the audiology field focus incorporate EHFA and DPOAEs specifically for this patient population.

### ***Monitoring Schedule***

Another challenge in the establishment of a clinical protocol would be determining how often audiological evaluations should be repeated. The general practice of audiologists and graduate students is to schedule annual hearing evaluations. Having an annual schedule may be an appropriate method considering that the patient may need to attend many other healthcare appointments in the context of monitoring their health.

A review of the literature also indicates a lack of evidence supporting fluctuating hearing loss associated with systemic lupus erythematosus (Di Stadio & Ralli, 2017; Rahne et al., 2017). Therefore, the nature of the hearing loss tends to be more progressive and unlikely for the thresholds to recover. As a result, the audiologist can anticipate a more standardized monitoring protocol that does not have to incorporate additional appointments in order to document the fluctuation.

Some respondents noted that they advise SLE patients to contact them for earlier testing if the patient notes subjective changes in hearing or concerns about their hearing arise before the routine annual test. In the case of sudden sensorineural hearing loss (SNHL), additional hearing evaluations may need to be scheduled in order to confirm the sudden hearing loss or to document the fluctuations in hearing

sensitivity. The research suggests that patients with SLE have a higher rate of sudden SNHL than individuals without the disease (Lin et al., 2013). Recent publications have identified even higher rates of sudden SNHL in patients with SLE who also have additional disorders, such as rheumatoid arthritis and antiphospholipid syndrome (Riera et al., 2019; Xie et al., 2019).

Considering the association of sudden SNHL, SLE, and the presence of comorbid diseases, the audiologist may need to have an active role in clinical team providing the patient care. This may require the audiologist to collaborate with the rheumatologist or primary care physician in order to determine the best monitoring schedule. The monitoring schedule may also be impacted by insurance recommendations and reimbursement or referral requirements as well.

### ***Professional Consensus***

A professional consensus is needed to generate an interdisciplinary clinical protocol. The best approach is to have a team of rheumatologists, otolaryngologists or otologists, and audiologists who have extensive experience in providing care to patients with SLE collaborate on a peer-reviewed guideline. This will permit a more finite step-by-step course of action and eliminate potential conflicting opinions regarding patient care. This document may also inform insurance companies with regard to coverage for audiological services.

### **Additional Resources**

In addition to the development of a protocol, there were other requests from the respondents to have a list of SLE support groups to provide to patients. Appendix E includes a handout to provide to patients who express interest in finding online and local resources. These resources also allow the audiologist to read patient stories and

become connected to other medical professionals that support patients with SLE. Appendix E provides a list of these groups, all of which have online resources. It is encouraged to have the audiologist become familiar with whether there are in-person resources within their general area, as the availability of this type of support varies depending on where the patient resides geographically. It should be noted that there is an opportunity for the protocol to recommend the audiologist to provide these resources and serve as a reminder to the audiologist of the availability of this information.

Furthermore, a brief list of relevant research articles has also been provided in Appendix E as a starting point for audiologists and graduate students to learn additional information. This is beneficial for audiologists who may have access to PubMed or an alternative database. Appendix E also provides a table of other online resources for an audiologist to become acquainted with SLE characteristics. A list of research articles to have in the clinic was requested by the majority of respondents and this appendix would also address the concern that audiologists and graduate students know which resources to utilize. While the references section of this research provides a comprehensive list of resources, this abbreviated list to have on hand in the clinic as a guide may be advantageous as a quick resource. These resources and protocol are to provide audiologists and audiology graduate students a set of materials so that they may adequate care to a group of patients that can vary in audiologic symptoms.

### **Accessibility to New Research**

A search of lupus on the frequently used resource for audiologist continuing education credits, AudiologyOnline.com, determined that there are no current publications or presentations regarding hearing loss and autoimmune diseases. On the

other hand, a search of diabetes found four links; two of which were a series of presentations outlining hearing loss in chronic disorders (which included diabetes). There may be a benefit in the development of a similar presentation: An informational compilation of autoimmune diseases associated with hearing loss that included SLE and conditions such as Wegener's granulomatosis, rheumatoid arthritis, Sjogren's disease, and Cogan syndrome. Again, taking into account that the SLE research is relatively new, an electronic continuing education presentation that was easily accessible would allow audiologists to keep up to date.

### **Initial Development of a Protocol**

Appendix F outlines a very general protocol that can provide the initial recommendations of audiologic care for a patient with SLE. Sections include what questions to incorporate into the case history, what baseline audiometric battery should be used, and what to keep in mind when determining a monitoring schedule. Again, additional research needs to be completed for the development of a formal protocol but the intention of this current protocol is to provide a temporary guideline for audiologists and graduate students.

### **Study Limitations**

A number of study limitations were identified throughout this research study. The majority of the respondents were enrolled in graduate school at the time they completed the survey. Potentially, a larger focus of practicing audiologists would have provided a more dynamic perspective of experiences of treating patients with SLE. Furthermore, the survey was distributed using an electronic link. As a result, the survey was not accessible to audiologists who rarely use technology during their personal time or do not have Facebook. It is anticipated that more respondents would

have been reached if letters with the survey link were handed out in person at audiology conferences.

Other limitations are in relation to the structure of the survey. Noted in the open comments section at the end of the survey, the concept of not providing an “I don’t know” option was mentioned. During the development of the survey, having this option was contemplated. It was concluded to have a closed set of questions in order to determine what most likely action they would take. Bearing in mind that this is a topic just being integrated into the field of audiology, providing an “I don’t know” option would not have captured what direction the respondent was leaning towards. As a result, it may have prevented participants from fully participating in the survey and leaving the survey incomplete. Additionally, the question “are you familiar with the autoimmune disease by the name of systemic lupus erythematosus?” may have deterred respondents who were not confident on the subject area. Ideally, the survey would have indicated “we are still interested in your survey responses.”

It would have been beneficial to clarify in the survey that SLE is commonly referred to as simply lupus. While there are multiple types of lupus, such as discoid lupus, there are higher rates of SLE and as a result is what is typically being referred to when the term lupus is used.

### **Future Research**

This research study is the first known study of a qualitative design addressing how audiologists and audiology graduate students provide care to patients with SLE. Additional research should identify which medical or audiological clinics have a clinical protocol and gain additional information regarding the actual administration of audiologic care to SLE patients. It may be feasible to mine the diagnostic

(international classification of diseases) and audiological procedure (current procedural terminology) codes submitted to third-party payers for a better sense of what care is being provided and the timeline for evaluations. Lastly, it might be informative to survey patients with SLE regarding their audiological care and experiences. Faculty surveys regarding audiology curriculums and course syllabi might also provide a clearer picture regarding the inclusion of SLE as an educational topic in graduate audiology training programs. The information provided in Appendices E and F is also beneficial for integration into coursework.

### **Conclusion**

This sample comprised of audiology graduate students and licensed audiologists has provided a snapshot of the general approaches these two groups are implementing in the audiologic care of patients with SLE. This study has identified that new and experienced audiologists need more information and resources in order to provide appropriate care. The current status of both groups is that there is a general awareness of the disease but a limited degree of confidence in providing care. These outcomes highlight the opportunities to develop a clinical and diagnostic protocol, to develop additional trainings, and the need to have this topic integrated into current graduate school courses. With the recent publications of SLE prevalence in the United States clarifying how patient denizens have this disease, it has become clearer that there is a likelihood that an audiologist will provide care to this population group during their career providing direct-patient care.

This study has underlined the opportunity for a group of professionals to work on a peer-reviewed clinical protocol. Further research should identify the appropriate actions an audiologist must complete in order to ensure that sufficient long-term

audiologic care can be provided to patients diagnosed with SLE. The need for standardized care and trained audiologists is critical for patients with SLE.

## REFERENCES

- Abbasi, M., Yazdi, Z., Kazemifar, A., & Bakhsh, Z. (2013). Hearing loss in patients with systemic lupus erythematosus. *Global Journal of Health Science*, 5(5), 102-106. doi:10.5539/gjhs.v5n5p102
- Akirav, E., Lebastchi, J., Galvan, E., Henegariu, O., Akirav, M., Ablamunits, V., Lizardi, P., & Herold, K. (2011). Detection of  $\beta$  cell death in diabetes using differentially methylated circulating DNA. *Proceedings of the National Academy of Sciences*, 108(47), 19018-19023. doi:10.1073/pnas.1111008108
- Al-Sukaini, A., Azam, M., & Samanta, A. (2014). Specific conditions. In *Rheumatology: A clinical handbook* (pp. 55-60). Scion Publishing Limited.
- Anderson, M., & Su, M. (2011). Aire and T cell development. *Current Opinion in Immunology*, 23, 198, 206. doi: 10.1016/j.coi.2010.11.007
- Andonopoulos, A., Naxakis, S., Goumas, P., & Lygatsikas, C. (1995). Sensorineural hearing disorders in systemic lupus erythematosus. *Clinical and Experimental Rheumatology*, 13(2), 137-141.
- Batuecas-Caletrío, A., Pino-Montes, J., Cordero-Civantos, C., Calle-Cabanillas, M., & Lopez-Escamez, J. (2013). Hearing and vestibular disorders in patients with systemic lupus erythematosus. *Lupus*, 22, 437-442. doi:10.1177/0961203313477223
- Bijl, M., & Kallenberg, C. (2006). Ultraviolet light and cutaneous lupus. *Lupus*, 15, 724-727. doi:10.1177/0961203306071705
- Bortoli, R., & Santiago, M. (2007). Chloroquine ototoxicity. *Clinical Rheumatology*, 26(11), 1809-1910. doi:10.1007/s10067-007-0662-6
- Botelho, C., Carvalho, S., & Novato, I. (2014). Increased prevalence of early cochlear damage in your patients with type 1 diabetes detected by distortion product otoacoustic emissions. *International Journal of Audiology*, 53, 402-408. doi:10.3109/14992027.2013.879341
- Bovo, R., Aimoni, C., & Martini, A. (2006). Immune-mediated inner ear disease. *Acta Oto-laryngology*, 126, 1012-1021. doi:101016/B978-0-12-384929-8.00056-3
- Chabner, D. (2013). *The language of medicine* (9th ed.). Saunders Elsevier.



- Chansky, P., & Werth, V. (2017). Accidental hydroxychloroquine overdose resulting in neurotoxic vestibulopathy. *BMJ Case Reports*. doi:10.1136/bcr-2016-218786
- Cordeschi, S., Salvinelli, F., & D'Ascanio, L. (2004). Sensorineural hearing impairment in systemic lupus erythematosus: Sudden or progressive? *Clinical and Experimental Rheumatology*, 22, 653-653.
- Costenbader, K., Karlson, E., Liang, M., & Mandl, L. (2002). Defining lupus cases for clinical studies: The Boston weighted criteria for the classification of systemic lupus erythematosus. *The Journal of Rheumatology*, 29(12), 2545-2550.
- Dall'Era, M., Cisternas, M., Snipes, K., Herrinton, L., Gordon, C., & Helmick, C. (2017). The incidence and prevalence of systemic lupus erythematosus in San Francisco county: The California lupus surveillance project. *Arthritis and Rheumatology*, 69(10), 1996-2005. doi: 10.1002/art.40191
- Davidson, A., & Diamond, B. (2001). Autoimmune diseases. *The New England Journal of Medicine*, 345(5), 340-350. doi:10.1056/NEJM200108023450506
- Davidson, A., & Diamond, B. (2014). General features of autoimmune disease. In I. Mackay & N. Rose (Eds.), *The autoimmune diseases* (5th ed., pp.19-36). Elsevier.
- Di Stadio, A., & Ralli, M. (2017). Systemic lupus erythematosus and hearing disorders: Literature review and meta-analysis of clinical and temporal bone findings. *Journal of International Medical Research*, 0(0), 1-11. doi:10.1177/0300060516688600
- Dragin, N., Le Panse, R., & Berrih-Aknin, S. (2017). Autoimmune disease predisposition: Aire « protects » men. *Médecine Sciences (Paris)*, 33(2), 169-175. doi:10.1051/medsci/20173302012
- Fleisher, T., Straus, S., & Bleesing, J. (2001). A genetic disorder of lymphocyte apoptosis involving the fas pathway: The autoimmune lymphoproliferative syndrome. *Current Allergy and Asthma Reports*, 1(6), 534-540. doi:10.1016/S0889-8561(01)00011-X
- Glatke, T., & Robinette, M. (2007). Otoacoustic emissions. In R. J. Roeser, M. Valente, & H. Hosford-Dunn (Eds.), *Audiology diagnosis* (2nd ed., pp. 478-494). Thieme Medical Publishers.
- Gomides, A., Rosario, E., Borges, H., Gomides, H., Padua, P., & Sampaio-Barros, P. (2007). Sensorineural dysacusis in patients with systemic lupus erythematosus. *Lupus*, 16, 987-990. doi:10.1177/0961203307084160

- Green, L., & Miller, E. (2001) Sudden sensorineural hearing loss as a first manifestation of systemic lupus erythematosus: Association with anti-cardiolipin antibodies. *Clinical Rheumatology*, 20, 220–222.
- Grodzicky, T., & Elkon, K. (2002). Apoptosis: A case where too much or too little can lead to autoimmunity. *Mount Sinai Journal of Medicine*, 69, 208-219.
- Helmick, C., Felson, D., Lawrence, R., Gabriel, S., Hirsh, R., Kwoh, C. K. Liang, M. H., Mayes, M., Merket, P., Pillemer, S. R., Reveille, J. D., & Stone, J. (2008). Estimates of the prevalence of arthritis and other rheumatic conditions in the United States: Part I. *Arthritis & Rheumatism*, 58(1), 15-25.  
doi:10.1002/art.23177
- Hisashi, K., Komune, S., & Taira, T. (1993). Anticardiolipin antibody-induced sudden profound sensorineural hearing loss. *American Journal of Otology*, 14, 275-277.
- Izmirly, P., Wan, I., Sahl, S., Buyon, J., Belmont, M., Salmon, J., Askanase, A., Bathon, J. M., Geraldino-Pardilla, L., Ali, Y., Gnzler, E. M. Putterman, C., & Parton, H. (2017). The incidence and prevalence of systemic lupus erythematosus in New York County (Manhattan), New York: The Manhattan lupus surveillance program. *Arthritis and Rheumatology*, 69(10), 2006-2017.  
doi:10.1002/art.40192
- Johansen, P., & Gran, J. (1998). Ototoxicity due to hydroxychloroquine: Report of two cases. *Clinical and Experimental Rheumatology*, 16(4), 472-474.
- Jung, T., Rhee, C., Lee, C., Park, Y., & Choi, D. (1993). Ototoxicity of salicylate, nonsteroidal anti-inflammatory drugs, and quinine. *Otolaryngology Clinic of North American*, 26, 791-810.
- Karabulut, H., Dagli, M., Ates, A., & Karaaslan, Y. (2010). Results for audiology and distortion product and transient evoked otoacoustic emissions in patients with system lupus erythematosus. *The Journal of Laryngology & Otology*, 124, 137-140. doi:10.1017/S0022215109991332
- Karatas, E., Onat, A., Durucu, C., Baglam, T., Kanlikama, M., Altunoren, O., & Buyukhatipoglu, H. (2007). Audiovestibular disturbance in patients with systemic lupus erythematosus. *Otolaryngology-Head and Neck Surgery*, 136, 82-86. doi:10.1016/j.otohns.2006.06.1255
- Kariya, S., Kaya, S., Hizli, O., Hizli, P., Nishizaki, K., Paparella, M. M., & Cureoglu, S. (2016). Cochlear histopathologic findings in patients with systemic lupus erythematosus: A human temporal bone study. *Otology & Neurology*, 37(5), 593-597. doi:10.1097/MAO.0000000000001017

- Kastanioudakis, I., Ziavra, N., Voulgari, P., Exarchakos, G., Skevas, A., & Drosos, A. (2002). Ear involvement in systemic lupus erythematosus patients: A comparative study. *Journal of Laryngology and Otology*, *116*(2), 103-107. doi:10.1258/0022215021910032
- Khalili, H., Dastan, F., & Dehghan, S. (2014). A case report of hearing loss post use of hydroxychloroquine in a HIV-infected patient. *Daru*, *22*(1), 20. doi:10.1186/2008-2231-22-20
- Kim, J., & Werth, V. (2014). Chapter 5: Malar Erythema. In Matucci-Cerinic, M., & Furst, D., & Florentino, D., *Skin manifestations in rheumatic disease* (p. 37). Springer Science+Business Media.
- Klareskog, L., Malmstrom, V., Lundberg, K., Padyukov, L., & Alfredsson, L. (2011). Smoking citrullination and genetic variability in the immune-pathogenesis of rheumatoid arthritis. *Seminars Immunology*, *23*, 92-98. doi:10.1016/j.smim.2011.01.014
- Larsen, M., Friis, M., Karlsen, C., Poulsen, S., Lund, S., & Qvortrup, K. (2015). Auto-inflammatory challenge of the endolymphatic sac-Cochlear damage measured by distortion product oto-acoustic emissions. *Acta Oto-Laryngologica*, *135*, 758-764. doi:10.3109/00016489.2015.1032433
- Lasso de la Vega, M., Villarreal, I., Moya, J., & Garcia-Berrocal, J. (2017). Extended high frequency audiometry can diagnose sub-clinic involvement in a seemingly normal hearing systemic lupus erythematosus population. *Acta Oto-Laryngologica*, *137*(2), 161-166. doi:10.1080/00016489.2016.1219049
- Lewis, M., & Botto, M. (2006) Complement deficiencies in humans and animals: links to autoimmunity, *Autoimmunity*, *39*, 367-378. doi:10.1080/0891690600739233
- Lim, S., Bayakly, A., Helmick, C., Gordon, C., Easley, K., & Drenkard, C. (2014). The incidence and prevalence of systemic lupus erythematosus, 2002-2004: The Georgia lupus registry. *Arthritis and Rheumatology*, *66*(2), 357-368. doi:10.1002/art.38239.
- Lin, C., Lin, S., Weng, S., & Lin, Y. (2013). Risk of sudden sensorineural hearing loss in patients with systemic lupus erythematosus: A population-based cohort study. *Audiology and Neurotology*, *18*(2), 95-100. doi: 10.1159/000345512
- Maciazczyk, K., Durko, T., Waszczykowska, E., Erkiert-Polguj, A., & Pajor, A. (2011). Auditory function in patients with systemic lupus erythematosus. *Auris Nasus Larynx*, *38*, 26-32. doi:10.1016/j.anl.2010.04.008
- Pettigrew, H., Teuber, S., & Gershwin, M. (2009). Clinical significance of complement deficiencies. *Annals of the New York Academy of Sciences*, *1173*, 108-123. doi:10.1111/j.1749-6632.2009.04633.x

- Rahne, T., Claud, F., Plontke, S., & Keyber, G. (2017). Prevalence of hearing impairment in patients with rheumatoid arthritis, granulomatosis with polyangiitis (GPA, Wegener's granulomatosis), systemic lupus erythematosus. *Clinical Rheumatology*, 36, 1501-1510. doi:10.1007/s10067-017-3651-4
- Riera, J., Maliandi, M., Musurana, J., & Cavallasca, J. (2019). Sudden sensorineural hearing loss in systemic lupus erythematosus and antiphospholipid syndrome: An update. *Current Rheumatology Reserves*. doi:10.2174/15733971156661910161 01741
- Roverano, S., Cassano, G., Paira, S., Chiavarini, J., Graf, C., Rico, L., & Heredia, C. (2006). Asymptomatic sensorineural hearing loss in patients with systemic lupus erythematosus. *Journal of Clinical Rheumatology*, 12(5), 217-220. doi:10.1097/01.rhu.0000242777.71604.69
- Rynes, R., & Park, A. (1993). Introduction to symposium on antimalarial therapy and lupus. *Lupus*, 2 (SI).
- Seckin, U., Ozoarn, K., Ikiniciogullari, A., Borman, P., & Bostan, E. (2000). Hydroxychloroquine ototoxicity in a patient with rheumatoid arthritis. *Rheumatology International*, 19(5), 203-204. doi:10.1007/s002960000054
- Silva, C., Avcin, T., & Brunner, H. (2012). Taxonomy for systemic lupus erythematosus with onset before adulthood. *Arthritis Care Research (Hoboken)*, 64(12), 1787-1793. doi:10.1002/acr.21757
- Smith, S. M. (2005). Qualtrics [Computer software]. Provo, UT. Retrieved from <http://www.qualtrics.com>.
- Smith-Bouvier, D. L., Divekar, A. R., Sasidhar, M., Du, S., Tiwari-Woodruff, S. K., King, J. K., Arnold, A. P., Singh, R. R., Voskuhl, R. R. (2008). A role for sex chromosome complement in the female bias in autoimmune disease. *The Journal Experimental Medicine*, 205, 1099-1108. doi:10.1084/jem/20070850
- Somers, E., Marder, W., Cagnoli, P., Lewis, E., DeGuire, P., Gordon, C., Helmick, C. G., Wang, L., Wing, J. J., Dhar, J. P. Leisent, J. Shaltis, D., & McCune, W. (2014). Population-based incidence and prevalence of systemic lupus erythematosus: The Michigan Lupus Epidemiology and Surveillance Program. *Arthritis Rheumatology*, 66(2), 369-378. doi: 10.1002/art.38238
- Sone, M., Schachern, P., Paparella, M., & Morizono, N. (1999). Study of systemic lupus erythematosus in temporal bones. *Annals of Otolaryngology, Rhinology, & Laryngology*, 108, 338-344. doi: 10.1177/000348949910800404
- Sperling, N., Tehrani, K., Liebling, A., & Ginzler, E. (1996). Aural symptoms and hearing loss in patients with lupus. *Otolaryngology-Head and Neck Surgery*, 118(6), 762-765.

- Suzuki, M., Krug, M., Cheng, K., Yazawa, Y., Bernsteain, J., & Yoo, T. (1997). Antibodies against inner-ear proteains, in the sear of patients with inner-ear diseases. *Journal of Otorhinolaryngology and Its Related Specialities*, 59, 10-17.
- Valiente, R., Roldan, A., Villarreal, I., & Garcia, J. (2016). Extended high-frequency audiometry (9,000-20,000 Hz). Usefulness in audiological diagnosis. *Acta Otorrinolaringology Espanola*, 67(1), 40-44. doi:10.1016/j.otorri.2015.02.002
- Xie, S., Ning, H., She, Y., Jing, Q., Jiang, Q., Zhang, Y., Mei, L., Feng, Y., & Wu, X. (2019). Effect of systemic lupus erythematosus and rheumatoid arthritis on sudden sensorineural hearing loss. *Laryngoscope*, doi:10.1002/lar.28455
- Yu, C., Gershwin, E., & Chang, C. (2014). Diagnostic criteria for systemic lupus erythematosus: A critical review. *Journal of Autoimmunity*, 48-49, 10-13. doi:10.1016/j.jaut.2014.01.004
- Zheng, J., Ren, T., Parthasarathi, A., & Nuttal, A. (2001). Quinine-induced alterations of electrically evoked otoacoustic emissions and cochlear potentials in guinea pigs. *Hearing Research*, 154(1-2), 124-134. doi:10.1016/S0378-5955(01)00229-5

APPENDIX A  
INSTITUTIONAL REVIEW BOARD APPROVAL



*Institutional Review Board*

DATE: December 21, 2018  
 TO: Carla White, B.A. [SEP]  
 FROM: University of Northern Colorado (UNCO) IRB

PROJECT TITLE: [1227193-1] Audiologists' Knowledge, Attitudes, and Practices Regarding Systemic Lupus Erythematosus Patients with a Hearing Loss

SUBMISSION TYPE: New Project

ACTION: APPROVAL/VERIFICATION OF EXEMPT STATUS

DECISION DATE: December 21, 2018

EXPIRATION DATE: December 21, 2022

Thank you for your submission of New Project materials for this project. The University of Northern Colorado (UNCO) IRB approves this project and verifies its status as EXEMPT according to federal IRB regulations.

**Carla -**

***Thank you for your patience with the UNC IRB process. Your application materials and protocols are clear, thorough and are verified/approved exempt.***

***Please update the IRB administrator contact to Nicole Morse as Sherry May retired last summer. Once that update is made you may begin participant recruitment and data collection. As noted in your application, please submit***

*any amendments/modifications that are warranted following your pilot testing.*

*Best wishes with your research and don't hesitate to contact me with any IRB-related questions or concerns.*

*Sincerely,*

*Dr. Megan Stellino*

We will retain a copy of this correspondence within our records for a duration of 4 years.

If you have any questions, please contact Nicole Morse at 970-351-1910 or [nicole.morse@unco.edu](mailto:nicole.morse@unco.edu). Please include your project title and reference number in all correspondence with this committee.

This letter has been electronically signed in accordance with all applicable regulations, and a copy is retained within University of Northern Colorado (UNCO) IRB's records.



APPENDIX B  
CONSENT FORM AND SURVEY

**Audiologists' Knowledge, Attitudes, and Practices Regarding  
Patients with SLE and Hearing Loss**

Start of Block: Research Consent Form



CONSENT FORM: HUMAN PARTICIPANTS IN RESEARCH

**Project title:** Audiologists' Knowledge, Attitudes, and Practices Regarding Patients with Systemic Lupus Erythematosus

**Researcher:** Carla White, B.A., AuD. Student

**Contact Information:** whit8237@bears.unco.edu or at 207-790-1348

Thank you for your interest in participating in this project! The purpose of this research study is to develop a clinical audiologic protocol, to address the gaps in knowledge of hearing loss and systemic lupus erythematosus within graduate training and CEU programs, and to address the gap in knowledge of audiologists performing monitoring services for this patient population. A qualitative survey will aid in the identification of methods and techniques used by audiologists for specific systemic lupus erythematosus patients and what the reasoning for the decisions made.

In the demographics section, you will not be asked to provide your name, allowing your responses to remain anonymous. The graduate student researcher and the research advisor will be the only individuals reviewing these responses. This survey will take approximately fifteen minutes to complete and is broken down into four sections: participant demographics, knowledge, attitudes, and practices. Minimal risks accompany the participation in this survey.

At the University of Northern Colorado, the institutional review board (IRB) has approved this project to be completed. It is your decision on whether you would like to participate in this study and complete the survey, which will be respected and not cause you to lose any benefits that are entitled to you. After reading and understanding the information provided above, please answer all questions in the four sections of the survey. Completing the survey will provide your permission as a participant, an electronic signature is not necessary. If any concerns arise regarding the selection and treatment you receive as a research participant in this study, the contact information for the IRB administrator Sherry May at the Office of Sponsored Programs is at Kepner Hall, University of Northern Colorado, Greeley, CO 80639: (970) 351-1910. Your participation is greatly appreciated!

**End of Block: Research Consent Form**

---

**Start of Block: Please Provide the Following Demographic Information**

Q-Intro What best describes your current role in the field of audiology?

- Licensed Audiologist (1)
- Graduate Student-Audiology (2)
- None of the above (3)

*Skip To: End of Survey If What best describes your current role in the field of audiology? = None of the above*

**End of Block: Please Provide the Following Demographic Information**

Q1.1S How many years have you completed in your graduate career?

- <1 year (1)
- 1 - (2)
- 2 - (3)
- 3 - (4)
- 4 - (5)
- 5 - (6)
- 6 - (7)
- >7 years (8)

Q1.2S Are you currently in an internship or externship position?

Yes (1)

No (2)

Q1.3S How many years of clinical experience have you accumulated? This includes experience at the educational institution, internships, and externships..

<1 year (1)

1 - (2)

2 - (3)

3 - (4)

4 - (5)

5 - (6)

6 - (7)

>8 years (8)

Q1.4S What state are you currently in while earning your Au.D degree?

▼ Alabama (1) ... I do not reside in the United States (53)

End of Block: Demographics: Graduate Student

---

Q1.1LA What is your terminal degree designation? Select all that apply.

MA (1)

MS (2)

Au.D. (3)

Ed.D. (4)

Ph.D. (5)

Sci.D. (6)

Other (please specify) (7) \_\_\_\_\_

Q1.2LA What state are you currently practicing in?

▼ Alabama (1) ... I do not reside in the United States (53)

Q1.3LA How many cumulative years have you been practicing as a licensed audiologist?

- <1 year (1)
- 1-2 years (2)
- 3-4 years (3)
- 5-10 years (4)
- 11-15 years (5)
- 16-20 years (6)
- 21-25 years (7)
- 26-30 years (8)
- 31-35 years (9)
- 36-40 years (10)
- 40+ years (11)

Q1.4LA In what setting are you currently employed?

- Academic (1)
- Consultant (2)
- Corporate audiology group practice (3)
- ENT, patient care (4)
- Government (eg. VA, IHS) (5)
- Graduate student (Au.D.) (6)

- Graduate student (Ph.D.) (7)
- Hospital (8)
- Manufacturer (9)
- Medical Clinic (10)
- Military (11)
- Private Practice (12)
- Retired (13)
- School/Educational (14)
- Speech and hearing clinic (15)
- Other (specify) (16) \_\_\_\_\_

Q1.5LA Have you provided direct patient care after receiving your degree in Audiology?

- Yes (1)
- No (2)

*Skip To: End of Survey If Have you provided direct patient care after receiving your degree in Audiology? = No*

Q1.6LA Have you provided direct patient care after receiving your degree in Audiology?

Yes

No

Q1.7LA How many cumulative years have you practiced in a medical setting?

<1 year (1)

1-2 years (2)

3-4 years (3)

5-10 years (4)

11-15 years (5)

16-20 years (6)

21-25 years (7)

26-30 years (8)

31-35 years (9)

36-40 years (10)

40+ years (11)

End of Block: Demographics: Licensed Audiologist

Start of Block: Knowledge of Systemic Lupus Erythematosus

Q2.1 Have you heard or read about the autoimmune disease called "systemic lupus erythematosus (SLE)?"

Yes (1)

No (2)

Display This Question:

If Have you heard or read about the autoimmune disease called "systemic lupus ... = Yes



Q2.2 Do you have a personal relationship with an individual with systemic lupus erythematosus?

- Yes (1)
- No (2)

Display This Question:

If Have you heard or read about the autoimmune disease called "systemic lupus ... = Yes

Q2.3 How did you FIRST become generally aware about the autoimmune disease systemic lupus erythematosus in?

- By providing audiological care to a patient with SLE (1)
- From an audiology colleague (2)
- from an electronic media/internet source (3)
- From a graduate school course (4)
- From a personal relationship with someone who has SLE (5)
- From another health care professional (6)
- Peer reviewed journal article (7)
- From print, books, newspaper, magazine (8)
- From television (9)
- Other (please specify) (10) \_\_\_\_\_

Display This Question:

If Have you heard or read about the autoimmune disease called "systemic lupus ... = Yes

Q2.4 Please estimate how long ago did you FIRST become generally aware of systemic lupus erythematosus?

- Unsure (1)
- <1 year ago (2)
- 1-2 years ago (3)
- 3-4 years ago (4)
- 4-5 years ago (5)

- 5-10 years ago (6)
- 11-15 years ago (7)
- 16-20 years ago (8)
- 20-25 years ago (9)
- 26-30 years ago (10)
- 31-35 years ago (11)
- 36- 40 years ago (12)
- 40> years ago (13)

Q2.5 How did you first become aware about HEARING LOSS associated with systemic lupus erythematosus?

- By providing audiological care to a patient with systemic lupus erythematosus (1)
- From an audiology colleague (2)
- From an electronic media/internet source (3)
- From a graduate school course (4)
- From a personal relationship with someone with systemic lupus erythematosus (5)
- From another health care professional (6)
- From a peer reviewed journal article(s) (8)
- From print, books, newspapers, magazine (9)
- From television (10)
- Other (please specify) (7) \_\_\_\_\_

*Display This Question:*

*If How did you first become aware about HEARING LOSS associated with systemic lupus erythematosus? =  
From a graduate school course*

Q2.6 Which graduate course? Please provide the name and a brief description of the course.

---

Q2.7 How susceptible is a patient with systemic lupus erythematosus to hearing loss?

- Low susceptibility (1)
- Low to moderate susceptibility (2)
- Moderate to high susceptibility (3)
- High susceptibility (4)

Q2.8 What anatomical structures of the auditory system may be susceptible to damage from SLE autoimmune responses? Select all that apply

- Auditory cortex (1)
- Auditory nerve (2)
- Basilar membrane (3)
- Cristae ampullaris of the vestibular system (4)
- Endolymphatic sac (5)
- Inner hair cells (6)
- Maculae of the vestibular system (7)
- Ossicular chain (8)
- Outer hair cells (9)
- Spiral ganglion cells (10)
- Stapedius and tensor tympani muscles (11)
- Stria vascularis (12)

Tympanic membrane (13)

Q2.9 What type of hearing loss is commonly associated with hearing loss from SLE autoimmune responses?

- Central (5)
- Conductive (1)
- Mixed (3)
- Neural (4)
- Sensory (2)
- None of the above (6)

*Display This Question:*

*If What type of hearing loss is commonly associated with hearing loss from SLE autoimmune responses? = Sensory*

Q2.10 For patients with SLE, the greatest degree of hearing loss is mostly likely observed within what frequency range?

- Only frequencies below 125 Hz (1)
- The lower frequencies, from 125 Hz to 1000 Hz (2)
- The mid-frequencies, from 1000 Hz to 4000 Hz (3)
- The higher frequencies, between 4000 Hz and 8000 Hz (4)
- None of the above (5)

Q2.11 For patients with SLE, does the hearing loss tend to be bilateral or unilateral?

- Bilateral (1)
- Unilateral (2)
- Equal prevalence of both (3)
- More so bilateral, but unilateral has been reported (4)
- None of the above (5)

Q2.12 Does the nature of the hearing loss in SLE patients tend to be progressive or stable over time?

- Stable (1)
- Progressive (2)
- The nature of the hearing loss is dependent on individual person's case of SLE (3)
- I don't know (4)

Q2.13 Do you believe that the duration of SLE and the degree of hearing loss are correlated? (e.g the longer a person has had SLE, the poorer the hearing).

- Yes (1)
- No (2)

Q2.14 Can systemic lupus erythematosus patients experience vertigo that is attributed to the presence of the autoimmune disease?

- Yes (1)
- No (2)
- Unsure (3)

Q2.15 Systemic lupus erythematosus is thought to arise in patients from which of the following factors:

- Genetics (1)
- Environmental factors (2)
- Both genetic and environmental factors (3)
- None of the above (4)

Q2.16 Based on your knowledge, which medications used in the treatment of systemic lupus erythematosus may be ototoxic? Select all that apply

- Prednisone (Corticosteroid) (2)
- Methotrexate (NSAID) (3)
- Hydroxychloroquine (Antimalarial) (1)
- Cyclophosphamide (Immunosuppressive) (4)

- Azathioprine (Immunosuppressive) (5)
- Mycophenolate (Immunosuppressive) (6)
- Belimumab (Immunosuppressive) (7)
- None of these medications are considered ototoxic (9)
- Other (please specify) (10) \_\_\_\_\_

End of Block: Knowledge of Systemic Lupus Erythematosus

---

Start of Block: Attitudes Towards Systemic Lupus Erythematosus

Q3.1 Do you think additional test procedures are necessary beyond the basic audiometric test battery (air conduction, bone conduction, speech recognition threshold, and word recognition in quiet) when evaluating patients with SLE?

Yes (1)

No (2)

*Display This Question:*

*If Do you think additional test procedures are necessary beyond the basic audiometric test battery (... = Yes*

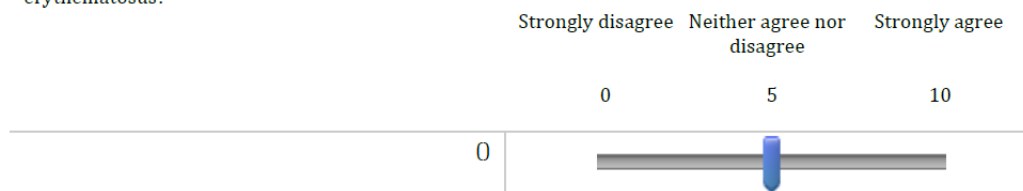
Q3.2 Which tests would you add to the test battery? Please select all that apply and a brief description of your reasoning.

- Acoustic Reflex Threshold (ART) (1)
- Acoustic Reflex Decay (2)
- Click-Evoked Auditory Brainstem Response (ABR) (3)
- Distortion Product Otoacoustic Emissions (DPOAEs) (4)
- Electrocochleography (ECoChG) (5)
- Extended High Frequency Audiometry (6)
- Extended High Frequency DPOAEs (7)
- Immittance Testing (8)
- Multifrequency Tympanometry (9)
- Transient Evoked Otoacoustic Emissions (TEOAEs) (10)
- Wide-Band Reflectance (11)
- Other (12) \_\_\_\_\_

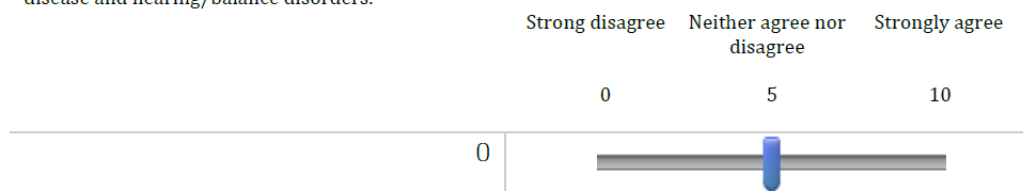


Q3.2 For the following questions, move the slider to report the extent in which you agree with the statement.

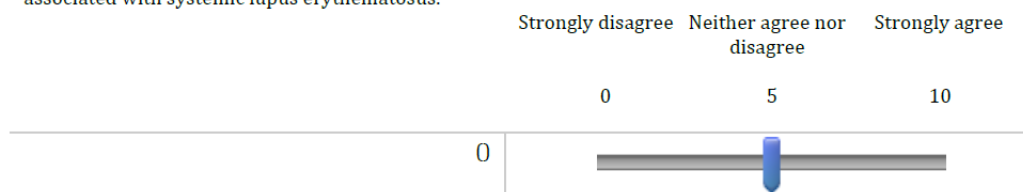
Q3.3-1 I am confident with my knowledge and understanding of the autoimmune disease systemic lupus erythematosus?



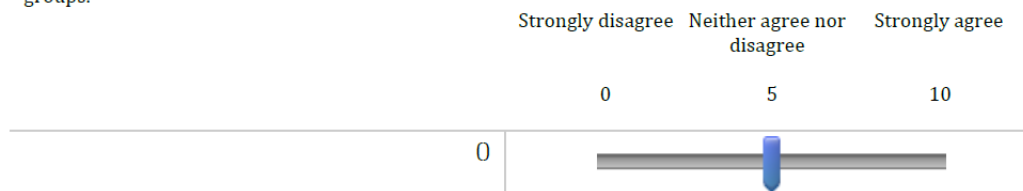
Q3.3-2 I feel confident when counseling systemic lupus erythematosus patients about autoimmune inner ear disease and hearing/balance disorders.



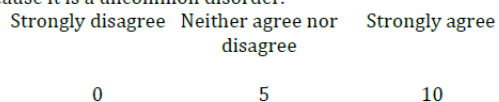
Q3.3-3 I have access to useful resources to find additional information on autoimmune inner ear disease associated with systemic lupus erythematosus.

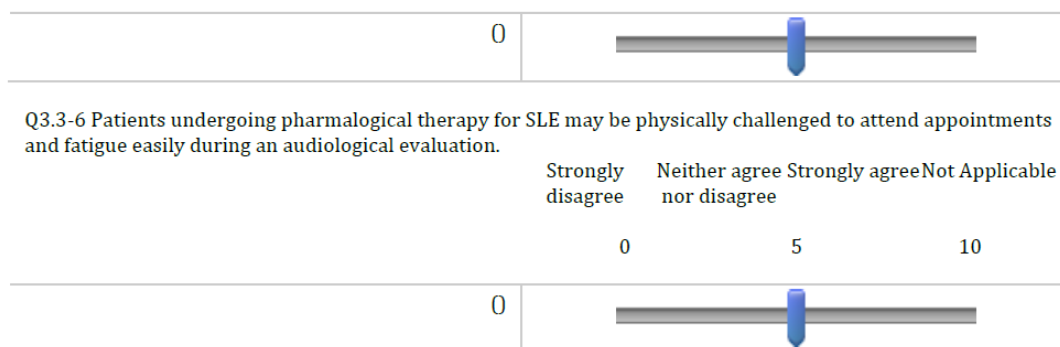


Q3.3-4 It is NOT important for patients with systemic lupus erythematosus to be referred to patient support groups.



Q3.3-5 Au.D. graduate programs should NOT be expected to provide academic and clinical training regarding autoimmune inner ear disease associated with SLE because it is a uncommon disorder.





End of Block: Attitudes Towards Systemic Lupus Erythematosus

Start of Block: Practices Associated with Systemic Lupus Erythematosus

Q4.1 Have you provided audiological care to a patient with systemic lupus erythematosus?

Yes (1)

No (2)

Q4.2 During your career, please estimate how many patients with systemic lupus erythematosus have you provided audiological care?

1-2 (1)

3-4 (2)

5-10 (3)

10-15 (4)

15-20 (5)

20-30 (6)

30-50 (7)

>50 (8)

Q4.3 How often do you recommend patients with systemic lupus erythematosus to have a hearing evaluation?

Annually (1)

Every 6 months (2)

Every 3 months (3)

Every month (4)

Other (5) \_\_\_\_\_

Q4.4 Do you typically include additional tests to the standard audiometric test battery for patients with SLE?

Yes (1)

No (2)

Q4.5 Please select all of the types of referrals you typically advise for a patient with SLE.

Family Practice Physician (1)

- Neurologist (2)
- Otolaryngologist (3)
- Otolologist (4)
- Rheumatologist (5)
- Other (please specify) (6) \_\_\_\_\_

Q4.6 Does your clinic or practice have a written clinical protocol for the provision audiological services to patients with systemic lupus erythematosus?

- Yes (1)
- No (2)

Display This Question:

*If Does your clinic or practice have a written clinical protocol for the provision audiological serv... = Yes*

Q4.7 Can we contact you for a copy of this protocol? If so, please provide your email address.

\_\_\_\_\_

Display This Question:

*If Does your clinic or practice have a written clinical protocol for the provision audiological serv... = No*

Q4.8 Would you be interested in implementing a clinical protocol for SLE patients if one was made available to you?

- Yes (1)
- No (2)

Display This Question:

*If Would you be interested in implementing a clinical protocol for SLE patients if one was made avai... = Yes*

Q4.9 What information would be useful to you in your clinical practice with regard to audiological care for patients with SLE Select all that apply.

- SLE resource list (1)
- List of interdisciplinary healthcare team members (2)

- Clinical diagnostic and monitoring protocol (3)
- Support group contacts (4)
- Other (please specify) (5) \_\_\_\_\_

Q4.10 Have you received additional training or pursued continuing education units (CEUs) on topics relating to autoimmune inner ear disease?

- Yes (1)
- No (2)

Q4.11 Do you have any other comments, experiences, or questions related to the provision of audiological care to SLE patients?

- Yes (1)
- No (2)

*Display This Question:*

*If Do you have any other comments, experiences, or questions related to the provision of audiological... = Yes*

Q4.12 Please use the text box below to leave any comments, experiences, or questions.

---



---



---



---



---

Closing Statement: Thank you for committing the time to complete this survey! If you have colleagues within the field of audiology that may provide valuable information on patients with systemic lupus erythematosus and hearing loss, or audiology colleagues that would like to participate in this research project, please feel free to forward the survey link to them.

**End of Block: Practices Associated with Systemic Lupus Erythematosus**

**Start of Block: Demographics: Graduate Student**

APPENDIX C  
CLINICAL RESEARCH ARTICLES RESOURCE

### **Prevalence of Hearing Loss and Audiologic Symptoms Associated with SLE**

- Karatas, E., Onat, A., Durucu, C., Baglam, T., Kanlikama, M., Altunoren, O., & Buyukhatipoglu. (2007). Audiovestibular disturbance in patients with systemic lupus erythematosus. *Otolaryngology-Head and Neck Surgery*, 136, 82-86. doi: 10.1016/j.otohns.2006.06.1255
- Kariya, S., Kaya, S., Hizli, O., Hizli, P., Nishizaki, K., ... & Cureoglu, S. (2016). Cochlear histopathologic findings in patients with systemic lupus erythematosus: a human temporal bone study. *Otology & Neurology*, 37(5), 593-597. doi: 10.1097/MAO.0000000000001017
- Rahne, T., Claud, F., Plontke, S., & Keyber, G. (2017). Prevalence of hearing impairment in patients with rheumatoid arthritis, granulomatosis with polyangiitis (GPA, Wegener's granulomatosis), systemic lupus erythematosus. *Clinical Rheumatology*, 36, 1501-1510. doi: 10.1007/s10067-017-3651-4

### **Diagnostic Monitoring**

- Karabulut, H., Dagli, M., Ates, A., & Karaaslan, Y. (2010). Results for audiology and distortion product and transient evoked otoacoustic emissions in patients with system lupus erythematosus. *The Journal of Laryngology & Otology*, 124, 137-140. doi: 10.1017/S0022215109991332
- Lasso de la Vega, M., Villarreal, I., Moya, J., Garcia-Berrocal, J. (2017). Extended high frequency audiometry can diagnose sub-clinic involvement in a seemingly normal hearing systemic lupus erythematosus population. *Acta Oto-Laryngologica*, 137(2), 161-166. doi: 10.1080/00016489.2016.1219049

### **Ototoxicity Treatment with Patients with SLE**

- Khalili, H., Dastan, F., & Dehghan, S. (2014). A case report of hearing loss post use of hydroxychloroquine in a HIV-infected patient. *Daru*, 22(1), 20. doi: 10.1186/2008-2231-22-20
- Seckin, U., Ozoarn, K., Ikiniciogullari, A., Borman, P., & Bostan, E. (2000). Hydroxychloroquine ototoxicity in a patient with rheumatoid arthritis. *Rheumatology International*, 19(5), 203-204. doi: 10.1007/s002960000054

APPENDIX D  
SURVEY KNOWLEDGE SECTION ANSWER KEY



Question	Correct answer
<p>How susceptible is a patient with systemic lupus erythematosus to hearing loss?</p> <p>What type of hearing loss is commonly associated with hearing loss from SLE autoimmune responses?</p> <p>For patients with SLE, does the hearing loss tend to be bilateral or unilateral?</p> <p>Do you believe that the duration of SLE and the degree of hearing loss are correlated? (e.g., the longer a person has had SLE, the poorer the hearing)</p> <p>Can SLE patients experience vertigo that is attributed to the presence of the autoimmune disease?</p> <p>SLE is thought to arise in patients from which of the following factors:</p> <p>The following medications are used in the treatment of SLE. Based on your knowledge, which ones can be ototoxic, if any? Selected choice.</p>	<p>Moderate to high susceptibility</p> <p>Sensory &amp; Neural</p> <p>More so bilateral, but unilateral has been reported</p> <p>Yes</p> <p>Yes</p> <p>Both genetic and environmental factors</p> <p>Hydroxychloroquine</p>

APPENDIX E

SYSTEMIC LUPUS ERYTHEMATOSUS PATIENT  
SUPPORT GROUP RESOURCE

Name of the Group	Description	Connections to Local Support Services?	Advocating Opportunities?
Lupus Foundation of America <a href="https://www.lupus.org/">https://www.lupus.org/</a>	The national lupus organization that has the mission of providing research updates, opportunities to join research trials, and educate the general public of the disease.	Yes	Yes
Molly's Fund Fighting Lupus <a href="https://kaleidoscopefightinglupus.org/">https://kaleidoscopefightinglupus.org/</a>	A nonprofit group that informs patients and professionals of the disease and oversees the development of government advocacy events.	No	Yes
Lupus Warriors Private Facebook Page	Provides patients and family members opportunities to connect with other lupus patients and pose general questions or concerns.	Yes	Yes
Daily Strength Lupus Group <a href="https://www.dailystrength.org/group/lupus">https://www.dailystrength.org/group/lupus</a>	Online blog to become connected with other lupus patients and family members	No	No

APPENDIX F  
CLINICAL PROTOCOL GUIDELINE

### Case History

1. Hearing Sensitivity Characteristics
  - a. Have you experienced a sudden change in hearing sensitivity in one ear or both ears?
  - b. When did you first notice a change in your hearing? Has it been slowly progressive or sudden?
2. Auditory Symptoms
  - a. Tinnitus
    - 1). Do you experience tinnitus in one ear or in both ears? If so, when did this first start?
    - 2). Over time, what has been your exposure to high-intensity sounds in occupational and recreational environments?
  - b. Vertigo
    - 1). If you experience any dizziness or imbalance, when does it occur? Does your dizziness coincide with flare symptoms?
3. Medications
  - a. What medications are you currently on?
  - b. What is an estimated amount of time you have been taking hydroxychloroquine (Plaquenil)?
4. Systemic Lupus Erythematosus Characteristics
  - a. Approximate time of diagnosis?
  - b. Comorbid disorders/diseases?
  - c. When was your last flare?
5. Identification of Medical Team
  - a. Rheumatologist?
  - b. Other professionals?

### Baseline Audiometric Battery

#### Recommended

1. Otoscopy
2. Tympanometry
3. Acoustic reflex testing
4. Distortion Product Otoacoustic Emissions (DPOAEs)
5. Pure tone audiometry (250-8000Hz) via air conduction and bone conduction
6. Extended high-frequency audiometry (EHFA) following SRO protocol
7. Speech recognition threshold (SRT) testing
8. Word recognition (WRS) testing

#### Case Dependent

1. If experiencing vertigo symptoms, possibly an VNG
2. Tinnitus matching protocol if tinnitus is reported

### Monitoring Schedule

1. Consider every six months if the patient has a comorbid autoimmune disease
2. Annual hearing with counseling that emphasizes that if a sudden hearing loss is perceived that immediate follow up with the primary care physician and rheumatologist is crucial
3. Consider contacting the rheumatologist to determine monitoring schedule

### Additional Considerations

1. Do additional referrals need to be made (ENT, otologist, neurologist)?
2. What releases of information need to be completed?