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**AUTOIMMUNE DISEASES AND VOICE:  
A CLINICAL PERSPECTIVE**



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# Autoimmune Diseases and Voice: Clinical Perspectives

**Mary Sandage, PhD, CCC-SLP**



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Over the many years I have treated voice disorders, it seems that a larger percentage of my current clinical caseload is comprised of individuals with voice changes secondary to autoimmune diseases. The nature of voice impairment secondary to autoimmune diseases is different than that for phonotraumatic lesions and muscle tension dysphonia, which

often require time-limited assessment and treatment for the disorder to achieve resolution. Atypical laryngeal findings, such as bamboo nodules, may also be a first clinical sign that an individual has an autoimmune disorder, as was the case for a recent young singer I cared for – observation of bamboo nodules led to the diagnosis of rheumatoid arthritis.

For this issue of the Newsletter, I asked Laura Dominguez, M.D. to provide the laryngologist's perspective on autoimmune disease and voice care. As Dr. Dominguez describes in her essay, AI disorders are chronic in nature, requiring us to keep in mind that these individuals may have recu-

rence of voice symptoms as they experience exacerbations of the disease process. I also asked two well-respected voice pathologists, Christine Kang and Sarah Schneider, to provide their perspectives for providing care for these individuals who present with intermittent voice difficulties.

Care for patients who have voice difficulty secondary to autoimmune disorders requires special consideration, particularly in light of the differences in clinical care required given the recurrence and remission nature of their voice difficulties and the ongoing nature of these clinical relationships that will be developed with these clients.

## Letter from the Editor

# Autoimmune Disease And Voice



**by Laura M. Dominguez, MD**

Autoimmune (AI) disease affects roughly 23.5 million Americans and is one of the leading causes of death in women.<sup>1</sup> Between 80-100 diseases have been identified thus far. Research continues into these complex disorders that generate nearly \$100 billion in annual health care costs. AI disorders are notorious for their effects on the musculoskeletal, renal, and dermatologic systems, amongst

many others. Many patients and physicians may not realize the larynx can also be affected and result in dysphonia and dyspnea. In some cases, these symptoms may precede manifestations in other organ systems. The Otolaryngology team may actually be the first to recognize the signs and symptoms of AI disease.

One of the most commonly encountered AI dis-

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- **SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)**

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- **RHEUMATOID ARTHRITIS (RA)**

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- **GRANULOMATOSIS POLYANGITIS (GPA)**

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orders is Systemic Lupus Erythematosus (SLE). SLE is a collagen vascular disease affecting a wide variety of organ systems. Most associate the classic “malar rash” with SLE and may not realize the larynx can also be affected. The laryngeal examination is key in identifying signs of SLE and other AI disorders in patients with voice complaints. The most common laryngeal symptoms in SLE patients are dysphonia and dyspnea, usually linked to edematous vocal folds. Ulcerations and even vocal fold paralysis can be present in certain cases. Edema (at the glottic and/or infraglottic level) and decreased vocal fold vibration can lead to severe hoarseness and strain. If the degree of edema is severe enough, this may result in a narrowed glottic airway, dyspnea, and stridor. The larynx is not commonly affected in SLE, but it is still critical to identify these manifestations early, particularly in cases of airway narrowing.

Rheumatoid arthritis (RA) is one of the other most commonly recognized AI disorders. While RA only affects a small percentage of the

general population, over 50% of those with the disease have laryngeal manifestations.<sup>2</sup> The most common are arthritis of the cricoarytenoid (CA) joint and rheumatoid lesions. CA joint arthritis can lead to reduced mobility of the vocal folds, dyspnea, and even stridor. Edema of the vocal folds in combination with reduced vocal fold mobility can lead to severe dysphonia in addition to respiratory complaints. If the patient does not have a previous diagnosis of AI disease, this presentation could be confused with vocal fold paralysis. It is important to distinguish between these conditions as treatment of the underlying RA can lead to resolution or improvement of hypomobility. Failure to recognize the true underlying condition, on the other hand, can lead to serious and life-threatening complications. There should be a high degree of suspicion in young female patients with a family history of AI disease. If there is a suspicion for AI disease lab work can be ordered. This usually includes antinuclear antibody (ANA) and rheumatoid factor (RF)

as SLE and RA are two of the most common disorders. Other labs such as antineutrophil cytoplasmic antibodies (c-ANCA/PR3) and anti-Ro (SSA) and anti-La (SSB) can be ordered if there is a suspicion for Granulomatosis with Polyangitis or Sjogren’s, respectively. A referral to rheumatology is placed for further testing and management.

Dysphonia can also relate to lesions of the vocal folds caused by AI disease. The most classic lesion is the “bamboo node”. This appears as a linear, cream-colored lesion extending across the mid-membranous true vocal fold. It can present as a single lesion or as a pair. Pathologically it represents submucosal edema with lymphocyte and neutrophil infiltration. The vibratory decrease that is associated with these lesions can result in severe dysphonia. It has been suggested these lesions are more common in women with heavy vocal demands. This is possibly related to phonotrauma and subsequent inflammation of microscopic vessels supplying the vocal folds. Resection

## ***“The Otolaryngology team may actually be the first to recognize the signs and symptoms of AI disease”.***

is controversial but generally avoided due to involvement of the vocal ligament and risk of permanent scarring. Typically, these lesions are treated with local steroid injections which can be done in the office setting or in the operating room. Voice therapy is also a viable option in this patient population. Management of the underlying AI disorder with disease modifying anti-rheumatic drugs (DMARDs) is ultimately the best method of treatment.<sup>3</sup>

Laryngeal lesions and edema are not the only way AI disease can affect the voice. The pulmonary system is intimately involved in phonation as the lungs are considered the “generator” of voice production. In the case of pulmonary involvement, which can present in a variety of AI disorders, projection and vocal quality can be compromised. One particular disease that can affect the respiratory tract signifi-

cantly is Granulomatosis Polyangiitis (GPA), formerly known as Wegener’s granulomatosis. This disease is linked to stenosis of the subglottis and trachea which can lead to respiratory difficulties. The disease can also result in pulmonary infiltrates and nodules which can lead to dyspnea and poor breath support. Once the airway has stenosed down to critical levels only surgical correction can improve the patient’s respiratory status. Treatment with DMARDs and corticosteroids is the ideal treatment to prevent disease progression and its manifestations in the respiratory tract.

One of the most important things to remember is that AI disorders are chronic in nature. Flares can occur resulting in recurrence of symptoms and fluctuating voice quality. In order to provide the best quality care, one needs to

recognize and appreciate the relationship between AI disease and the organ systems that affect voice production.

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by CHRISTINA KANG, MM, MS, CCC-SLP

# Autoimmune Diseases and Voice



**50M**

**Autoimmune Related Diseases Association projects an increase of 50 million in prevalence and types of autoimmune diseases.**

**The National Institute of Health projects an increase of 23.5 million in prevalence and types of autoimmune diseases;**

**23.5M**

I cannot say objectively that I have seen autoimmune diseases on the rise among voice patients in the last decade, but I can say with certainty that it is common, and that it is always a consideration when assessing patients.

Others have certainly noticed an increase. Both the American Autoimmune Related Diseases Association and National Institute of Health project differing numbers (50 million versus 23.5 million) in prevalence and types of autoimmune diseases (100 versus 80) but agree that autoim-

mune diseases are on the rise. Dr. Douglas Kerr of Johns Hopkins Hospital is a neurologist and neuroscientist who has penned a forward to Donna Nakazawa's *"The Autoimmune Epidemic."* He states that autoimmune diseases have not always been common but their prevalence has increased three times more than they were several decades ago. He points out that the increase in the prevalence is not due to increased recognition of these disorders or altered diagnostic criteria, but rather more people

are getting autoimmune diseases than ever before.

Autoimmune diseases are hard to diagnose because many symptoms overlap with symptoms of other conditions. Environmental change is often blamed since genes do not typically change in such a short period of time. In practice, factors such as genetics, environment, diet, and stress are well known in developing autoimmune diseases and must be taken into consideration. To further complicate things, a person



**“Autoimmune disease flare-ups exacerbate voice problems, and many patients have circulating autoantibodies acting against their own tissue, but have not yet been diagnosed with autoimmune diseases.”**

may have more than one autoimmune disease at the same time.

Autoimmune disease occurs when the body’s immune system does not distinguish between healthy tissue and antigens and mistakenly attacks healthy tissue. Some of the effects include destruction of body tissue, abnormal growth of an organ, and changes in organ function. Autoimmune diseases may affect one or more organ or tissue types including blood vessels, connective tissues, endocrine glands, joints, muscles, red blood cells and skin. Symptoms of autoimmune diseases vary based on the type and location of the faulty immune response. The common symptoms are fatigue, fever, inflammation, general malaise, joint pain, and rash. Treatment goals are 1) relieving/reducing symptoms, 2) controlling the autoimmune process, 3) maintain the body’s ability to fight the disease. Treatment depends on disease and symptoms such as immune suppression, tumor

necrosis factor blockers for some diseases, replacing vital substances that the body no longer makes in the form of supplements, blood transfusion if blood is affected, and physical therapy targeting movement if bones, joints, or muscles are affected.

Central sensitization syndrome (CSS) such as fibromyalgia also shares overlapping symptoms with autoimmune diseases such as fatigue, mood and cognitive problems, sleep disturbances, and multisensory hypersensitivity. It is possible to have CSS and autoimmune diseases at the same time. Patients may benefit from pain rehabilitation program.

In a multidisciplinary voice evaluation, our curiosity peaks when a patient complains of vague and disturbing symptoms such as excessive fatigue, brain fog or physical pain without any definitive diagnosis. Autoimmune disease flare-ups exacerbate voice problems, and many patients have circulating autoantibodies acting against their own tissue,

but have not yet been diagnosed with autoimmune diseases.

Women are more susceptible to both voice disorders and autoimmune diseases. An unpublished study that I conducted looking at the prevalence of voice disorders in patients with autoimmune diseases in all three Mayo Clinic sites from 2000-2010 revealed a sex bias of more than 65% of patients being female.

What my patients with autoimmune diseases have shown me over the years is the importance of mind-body intuition. I have observed 4 categories of patients 1) those who resign to managing their symptoms with medication and steroids, 2) those who swear by stress management to keep symptoms at bay and avoid flare ups, 3) those that swear by dropping all processed foods, sugars, white flour, caffeine and/or alcohol, and 4) those who pursue a balance of the previous options. . There is no evidence that diet will protect you from developing an autoimmune disease and such

**“In a multidisciplinary voice evaluation, our curiosity peaks when a patient complains of vague and disturbing symptoms such as excessive fatigue, brain fog or physical pain without any definitive diagnosis.”**

a belief risks “blaming the victim” mentality. But I also cannot deny that a poor diet and unmanaged stress generally tend to make things worse.

I find it beneficial for patients to understand the nature of inflammation. The word inflammation is often mentioned in the autoimmune disease context but what it means to one person may not be salient to another. As a simple example, when a person eats junk food and suffers

the symptom of acid reflux, that is an inflammation response caused by a behavior that the person can identify and alter. However it is easier to take medication and repeat the offending behavior. “Take a pill and you can eat another spicy chili hot dog drenched with processed cheese!” Sarcasm aside, words and thoughts can be inflaming too, since they are often connected to stress. We live in a time that promotes and rewards hyperfunction, overachievement, perfectionism, and making easy cuts for fast results. Being tired is the norm. Taking a different approach can have benefits. For example, in Yoga philosophy, one is encouraged to practice nonviolence in gestures, thoughts, and words, both to oneself and to others. While it is not necessary to become a yogi, in my observation, patients who have learned to adapt their behaviors and reduce stress by being mindful of their body-mind connection show the highest success in management of their autoimmune diseases.

One prevailing grievance often expressed by patients with autoimmune diseases is “not being taken seriously” by medical professionals, family, and society at large. Many had to be their own advocate to receive a diagnosis and treatment. A patient once told me in tears that her provider said that she was a hypochondriac and referred her to a psychiatrist. As providers we cannot put the burden on the patient just because the answer is not obvious. Empathy and patience are always appreciated.

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# autoimmune on the rise?

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# Considerations for Voice Therapy in Patients with Autoimmune Disease

**Sarah L. Schneider, MS, CCC-SLP**



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When treating a voice disorder there are several components that guide the speech-language pathologist's decision making. These components may include a thorough understanding of the case including onset and duration of symptoms, medical and surgical history, voice use patterns and vocal demands, perceptual evaluation, laryngeal palpation, acoustic and aerodynamic measures, and the laryngeal examination with videostroboscopy. As clinicians, we must put all of this information into

the context of what is happening with the patient related to their voice. And more holistically, we must consider what is happening in the patient's life that could be impacting their voice. It can be easy to evaluate laryngeal appearance and function, assess vibratory characteristics of the vocal folds, and develop a treatment plan, however, the larynx cannot be looked at in isolation. While there is growing knowledge regarding autoimmune disease, much is still unknown about these processes and how they may impact the voice. There is currently no gold standard for care of people with autoimmune disease and voice complaints (Todic et al., 2018). Resources to guide treatment include clinical experience and expert opinion, case reports, and a few retrospective case series. The current literature recommends voice therapy as the first line treatment for vocal fold lesions that are thought to be autoimmune related; however, voice therapy may also be combined with introduction or adjustment of systemic autoimmune medication, use of systemic or intralesional steroids, or even surgical excision<sup>1,2</sup> (Todic et al., 2018, Oker et al., 2019). As

***“As clinicians, we must consider all of these factors and meet the patient where they are, in their process, to address vocal needs.”***

clinicians, we must consider all of these factors and meet the patient where they are, in their process, to address vocal needs.

Autoimmune diseases may include systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), or Sjögren syndrome (SS) to name a few. Symptoms may manifest in hoarseness, stridor, running out of breath with talking, pain with speaking, increased vocal effort, reduced vocal intensity, and dysphagia. During laryngeal examination, depending on the autoimmune disease process, we may see changes in vocal fold mobility, laryngeal edema, or midmembranous vocal fold lesions. The lesions may be ‘bamboo’ nodules or rheumatoid nodules. Bamboo nodules are typically bilateral lesions with a yellowish/creamy appearance, located at the mid-membranous vocal

and with reduced vibration and incomplete vocal fold closure.

While large tertiary specialty centers may see more patients with autoimmune disease, it remains relatively uncommon. Bamboo nodules, while distinct in appearance, may be mistaken for traditional vocal fold nodules. Laryngeal examination with stroboscopy is necessary to help differentiate the vibratory characteristics of these vocal fold lesions. Bamboo nodules can sometimes be the first manifestation of an autoimmune disease and are often observed in middle aged women with high vocal demand. Considering this information, clinicians must be astute when completing case review and physical examination to pay attention to clues indicating possible autoimmune involvement. The multidisciplinary care team, including primary care, rheumatology, laryngology, and speech-language pathology,

is key for providing the highest level of care for patients with autoimmune disease and voice complaints. When voice changes are observed, referral to a laryngologist and voice trained speech pathologist should be made to further parse out these details and determine the most appropriate plan of care. While voice therapy may be the supported first line treatment for voice complaints in this population, the identification and management of the autoimmune disease is paramount.

Vocal fold nodules are the result of mechanical stress during vocal fold vibration. It is well accepted that voice therapy is the first-line, and often only, treatment and will result in improvement and even resolution of the lesions. **In the case of the bamboo nodules, it is thought that the disease process in combination with the mechanical stress of phonation contribute to their formation. While vocal symptoms may improve and lesions appear smaller, they may not resolve with therapy alone. They may require a combination of behavioral and medical therapies to maximize treatment outcomes and, even so, some voice**



**complaints may persist<sup>1,2</sup> (Todic et al., 2018, Oker et al., 2019).**

**Knowing this can provide insight for patient counseling and to help create realistic expectations regarding voice therapy outcomes.**

Patients often report variations in their vocal complaints related to a ‘flare-up’ or increased activity of autoimmune symptoms. This makes sense in that systemic autoimmune disease is not static. There is limited evidence in this area related to voice, possibly because other physical complaints outweigh the impact of voice changes and they are under-reported. While, understanding there may be variability in symptoms does not tell us how to treat the patient, this additional informa-

tion gives us context to validate patient complaints. Voice therapy may be completed during a flare-up; however, we may also exercise patience to let the flare-up of systemic symptoms calm down and address residual vocal symptoms as needed. Timing of voice therapy should be considered case by case based on the patient’s vocal complaints and demands.

While indirect and direct voice therapy techniques used with this patient population will likely not vary from traditional voice therapy, we have to understand how the vocal folds are functioning and the way the patient is using their voice to identify the most effective techniques for treatment to reduce mechanical stress of vocal fold vibration and improve vocal efficiency. And, with that, knowing the medical diagnosis and possible variability of systemic symptoms, we may have to adjust our expectations of therapy outcomes. Ideally, the patient’s complaints will improve or even resolve whether or not vocal fold lesions persist. However, we must consider, given the systemic disease and plethora of unknown variables that require more research to elucidate, that the voice may not return to normal but improve

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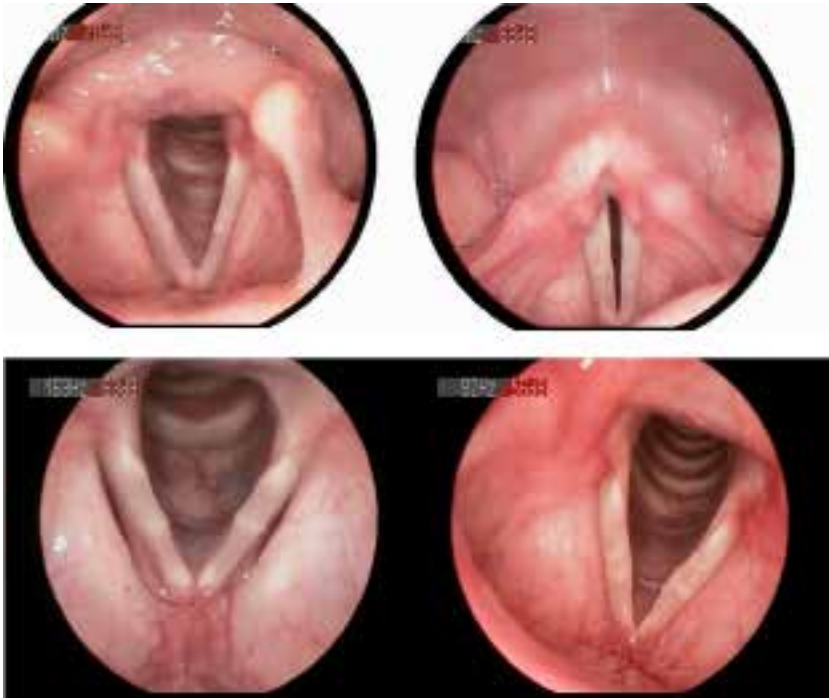


Figure 1. Examples of bamboo nodes under rigid indirect laryngoscopy: mainly bilateral, creamy-yellow, transverse, band-like deposits in the middle third of the vocal folds.

Journal of Voice

*Bamboo Nodes on a Series of 15 Patients: Vocal Fold Lesion as a Sign of Autoimmune Disease and Microphonotrauma*

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to a new baseline. As clinicians, we work to meet patients where they are in their process and, as a result, may maintain long term relationships with some patients acting as a support system for recalibrating voice production patterns and expectations through the process of their disease.

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