Neurological Problems of the Voice

What are neurological problems of the voice?

Normal speech production requires precise control and coordination of the muscles of the voice box, throat, palate, jaw, tongue, and lips. When the vocal fold muscles are dysfunctional, we generally refer to this symptom as a voice abnormality. On the other hand, when the tongue and lip muscles malfunction, the articulation of words becomes altered (typically "slurred"), resulting in a speech disorder. Difficulty forming words is a neurological problem of speech and is called dysarthria, a condition commonly seen after a stroke.

Neurological voice disorders are due to abnormalities of the brain and/or the nerves of the body that impair control of the muscles of the voice box, throat, palate, jaw, tongue, or lips, resulting in a variety of voice and/or speech problems. When neurological voice disorders lead to weakness of the vocal fold muscles, the voice may be very weak, breathy, and subject to fatigue. At other times a harsh and strained voice may be present due to vocal fold spasms from an underlying neurological condition. The voice may also sound too nasal (from weakness of the palate muscles), or speech patterns may be slurred or monotonous from tongue/jaw/lip involvement. The type of problem varies with the underlying disease process and the specific muscles and actions that are affected.

What are the symptoms of neurological voice disorders?

Isolated hoarseness is usually due to a problem within the larynx, but could be a sign of neurologic disease in some cases. Neurologic hoarseness is usually accompanied by other symptoms, such as speech distortion, swallowing problems, choking when drinking liquids, or neurological problems elsewhere in the body (i.e., arm or leg weakness or tremor). Acquired speech impairment (difficulty forming words) in adults virtually always indicates a neurologic problem. Poor control of the muscles that shape words is called dysarthria. This may range from slurring of speech to imprecise consonants, or completely incomprehensible speech. Dysfluent speech, such as stuttering or stammering, also indicates a neurological problem. Some patients know what they want to say, but cannot get the words out (aphasia). Other patients cannot remember words. These problems are usually the result of a stroke.

A change in resonance, such as an overly nasal sounding voice, often indicates impaired control of throat or palate muscles. A shaking, tremulous
voice indicates a tremor, which is seen in a variety of neurological disorders. Abrupt voice spasms are also signs of neurological dysfunction, although sometimes patients with emotional problems could have similar changes in the sound of the voice.

Difficulty breathing can be a sign of an underlying neurological disease and may be due to weakness/paralysis of both vocal folds resulting in a narrow, compromised airway.

**What are some of the more common neurological diseases that affect the voice?**

Because of the complex and diverse nature of neurological voice disorders, the more common conditions will be covered individually, addressing the following areas:

What is the cause of the disease and who is at risk?

What are the symptoms?

How is the disease diagnosed?

What are the pitfalls of diagnosis/common misdiagnoses?

How is the disease treated and what is the overall prognosis?

1. Stroke (Cerebrovascular Accident or CVA)
   - What is the cause of the disease and who is at risk?
     Stroke results from interrupted blood flow to the brain or brainstem, and is generally seen in males above the age of fifty. The risk factors for stroke include high blood pressure, high cholesterol level, and vascular disease.
   - What are the symptoms?
     Vocal symptoms typically consist of a hoarse, breathy voice when the brainstem is involved in stroke, and is due to paralysis of the vocal fold muscles on one side. Often, the patient also will experience swallowing difficulties as well as coughing/choking (especially when drinking liquids) that can be quite severe and debilitating. The combination of a paralyzed vocal fold, poor airway protection, weakened swallowing muscles, and loss of feeling/sensation in the throat probably explains the severe nature of swallowing problems in some stroke patients. Occasionally, a weak palate will cause a hypernasal voice as well.
   - Vocal symptoms with cerebral cortex strokes more often are related to articulation disorders (slurred speech, or dysarthria), rather than hoarseness. Some patients may have loss of speaking ability (aphasia) when the dominant brain hemisphere is affected.
   - How is stroke diagnosed?
     The diagnosis is made clinically (history and physical examination)
   - How is stroke treated and what is the prognosis?
     New clot-dissolving drugs, if given early during the emergence of stroke symptoms, can make a huge difference in the prognosis of stroke. The overall prognosis of stroke varies considerably, but in general, the older the patient the worse the prognosis for recovery of function. Vocal and swallowing difficulties often persist, and can be so severe that the patient is dependant on a feeding tube for nutrition and is unable to communicate. Speech and swallowing therapy with a Speech/Language Pathologist is essential in the rehabilitation process for many stroke patients.

2. Parkinson’s Disease
   - What are the symptoms?
   Voice symptoms consist of low volume voice with a monotone (expressionless) quality. The speech pattern is often produced in short bursts with inappropriate silences between words, and long pauses before initiating speech. The speech may be slurred as well. A small percentage of patients (about fifteen percent) may also have a tremulous voice. As with most neurological disorders, voice and speech disturbance are merely a small part of the symptoms in Parkinson’s. Tremor in the hands, and slow, shuffling gait, and other movement disturbances dominate, and often present long before vocal difficulties enter the picture.
   - What is the cause of the disease and who is at risk?
     The cause is not fully understood, but loss of nerve cells in select areas of the brainstem occurs through an unknown mechanism. The disease is more common in men (3:2 male to female ratio), and the age of onset is usually above the age of fifty. No risk factors are known.
   - How is it diagnosed?
     The diagnosis is made clinically (history and physical examination)
by a neurologist, although MRI or CAT scans are often obtained to help rule out other conditions.

- **What are possible misdiagnoses?**
  Exposure to certain toxic substances, such as carbon monoxide, and side effects from certain neuroleptic medications can cause symptoms similar to Parkinson’s disease.

- **How is Parkinson’s disease treated?**
  A team approach with involvement of Neurology, Speech Pathology, and Otolaryngology is recommended. Medication (Levodopa) is commonly used for the motor manifestations of Parkinson’s, but has little effect on the voice and speech. Rehabilitation of speech, voice, and swallow function is typically handled by a speech pathologist. The mainstay of the treatment of the communication problems (slurred speech and soft, weak voice) in patients with Parkinson’s disease involves the use of a specialized voice therapy treatment method called Lee Silverman Voice Treatment (LSVT®). Prior to LSVT®, speech therapy was used with very little success in patients with Parkinson’s disease; however, recent research has shown significant improvement in quality of life, speech, and swallow function following the LSVT® program. An otolaryngologist may be consulted to offer surgical treatments to help restore volume to the patient’s voice. Typically medialization laryngoplasty or injection laryngoplasty (with fat, collagen, and a variety of other substances) is used to bulk up the weakened vocal folds. Results of these procedures are often disappointing due to the global nature of the voice problem.

Deep brain stimulation has been used to treat Parkinson’s disease; however, voice symptoms may not improve, and may actually worsen with this treatment.

### 3. Spasmodic Dysphonia

- **What are the symptoms?**
  The symptoms of this neurological disorder are unique in that voice problems are most often the only symptom of the disorder. There are two different types of SD: adductor spasmodic dysphonia and abductor spasmodic dysphonia. Adductor SD is most common (eighty percent) and is characterized by a tight, strained, strangled voice caused by spasms of the muscles that move the vocal folds to the center of the voice box for producing voice. Abductor SD causes a breathy, weak voice during speech due to spasms of the muscles responsible for moving the vocal folds laterally (opening of the larynx for breathing). Often patients with SD will have a normal voice for simple sounds (saying “ah”), but voice breaks or spasms occur when more complex speech is attempted. Patients with SD also experience an increase in symptoms when there is an increase in stress. Some individuals have mixed spasmodic dysphonia, which involves spasms of both sets of muscles.

- **What is the cause of the disease and who is at risk?**
  Spasmodic dysphonia is a neurological disorder that is a dystonia of the muscles of the larynx. A dystonia is a neurological condition that causes spasms or spastic contractions of specific muscles when they are used. Dystonia can occur in a specific set of muscles (e.g., eyes, larynx, or hand), and when this occurs, it is called a focal dystonia. Dystonia can also occur to a region of the body (head and neck or arm) or to all the muscles of the body. The latter is very rare. Spasmodic dysphonia also has been called spastic dysphonia in the past. This condition is caused by an abnormality in the part of the brain (basal ganglia) responsible for control of laryngeal muscle contraction.

According to our best knowledge, spasmodic dysphonia usually is not a disease of heredity. Much research is being done regarding the genetic causes and links to the various dystonias. Recent research has found genetic causes, though, of other types of neurological movement disorders. It is hoped that this research will result in improved detection and treatment methods.

- **How is SD diagnosed?**
  An otolaryngologist with specialized interest and/or training in voice disorders is best suited to evaluate a patient with SD. It has been said that a trained ear is one of the best ways to diagnose SD, but other evaluations help solidify the diagnosis. Careful examination with fiberoptic laryngoscopy can reveal typical spasms of the vocal fold muscles during speech. In addition, voice spectography can be used to identify a characteristic striated pattern. Evaluation and voice therapy by a speech pathologist is sometimes attempted. Because SD is a neurological disorder, and not a functional one, voice therapy is often frustrating for patients, and ultimately ineffective.

- **What are the pitfalls of diagnosis?**
  Spasmodic dysphonia is often misdiagnosed by primary care physicians and otolaryngologists
alike. Because it is a relatively rare condition, even otolaryngologists may need to be familiar with the characteristic voice of a patient with SD, and misdiagnosis of the cause of hoarseness is often attributed to voice overuse and/or abuse, laryngopharyngeal reflux, muscular tension dysphonia, or may even be dismissed as functional/psychogenic. It is important for patients who believe they may have SD to seek the opinion of a voice specialist/laryngologist or speech pathologist with expertise in voice disorders.

• How is it treated and what is the prognosis?

In general, neurologic dysfunction cannot be cured, and treatment is aimed at reducing symptoms or assisting with adaptive strategies. This also is true with spasmodic dysphonia. The basic problem is impaired brain control of laryngeal muscles, resulting in uncontrolled laryngeal muscle spasms. But the only effective treatments are those aimed at the laryngeal muscle function. Local injections of botulinum toxin can weaken the involved muscles, so that spasms are reduced or eliminated. The underlying brain dysfunction cannot be corrected.

Botulinum toxin (Botox) injections are generally very successful for the treatment of spasmodic dysphonia. Patients with adductor SD are treated with Botox injection into the muscle(s) responsible for moving the vocal fold(s) to the center of the voice box (adduction) during phonation (thyroarytenoid and lateral cricoarytenoid muscles). For abductor SD, Botox is injected in the muscle(s) (posterior cricoarytenoid muscle) that retract the vocal folds (abduction). Both of these injections are usually done in the office with only local anesthesia (lidocaine placed under the skin of the injection site). The Botox injection for adductor SD is done 1 cm. below the Adam's apple with the patient lying flat or at a 45-degree angle. For abductor SD, the injection is done at the site of the voice box, and often the doctor will gently grasp and rotate the voice box to allow the injection needle to reach the intended muscle(s). Botox injections are usually done by laryngologists or ear, nose, and throat doctors with special training. The injections are often done with the assistance of electromyography (EMG). EMG records the electrical activity of muscle contraction and helps pinpoint the dysfunctional muscles. Botox is not active until a minimum of thirty-six hours after injection and causes a weakness of the muscles. This weakness reduces or eliminates the muscle spasms or over-contractures of SD. The effect of Botox is only temporary, and a typical injection to the laryngeal muscles lasts three months. Sometimes the Botox works longer and sometimes shorter, largely attributable to the dose and to factors not well understood by doctors.

4. Benign Essential Tremor (BET)

• What is BET?

BET is a neurologic disease that involves abnormal control of muscle contraction. The result is a tremor or shake of the involved muscle(s) either at rest or when in use. This often causes a periodic shake in the hand with and/or without use. BET can occur in the muscles of the palate, tongue, throat, and vocal folds. The result is an inability to keep the voice steady and thus the sound of the voice has periodic (regular) wobble or shake.

• What are the symptoms?

BET is one of the more common neurological disorders that can affect the voice. The three symptoms that may be present are tremors in the hands, head, and tremulous voice. All three may be present, or only one symptom may dominate.

• What is the cause of the disease and who is at risk?

As with most neurological disorders, the cause is not fully understood. BET is more common in females and often does not present until age forty to fifty. It may be inherited in certain instances. No other risk factors are known.

• How is it diagnosed?

Again, the diagnosis is most often a clinical one made by careful history and examination by a neurologist.

• What are the pitfalls of diagnosis?

As neurologists are not able to examine the larynx, BET may be misdiagnosed by a neurologist if only vocal tremors are present and the accompanying head and hand tremors are absent. In these cases, a fiberoptic laryngoscopy exam by a laryngologist/voice specialist may be needed to make the diagnosis. Other neurological disorders may have tremor as a component and be mistaken for BET, especially in the case of adductor SD with tremor. Other disorders such as Parkinson's disease, ALS, and cerebellar ataxia may have vocal tremor as a component of the voice disorder, but they are rarely mistaken for BET.
• How is it treated?
Medications are sometimes employed to control the tremor, such as propranolol; however, they usually have only a mild effect on the vocal tremor. Botulinum toxin injection into the vocal folds may be attempted, but success is generally lower than that for SD (usually fifty to sixty-five percent).

5. Amyotrophic Lateral Sclerosis (ALS, “Lou Gehrig’s” Disease)
• What are the symptoms?
ALS is a progressive neurological disease that may present initially with speech/voice difficulties as the primary symptoms in up to ten to fifteen percent of patients. These symptoms are spastic/strained voice, slurred speech, and hypernasal voice. Eventually other symptoms such as weakness and spasms of the limbs will appear (and predominate) in these cases. Most patients, however, notice weakness or incoordination in their arms or legs before they begin to experience vocal changes.
• What is the cause of the disease and who is at risk?
The cause is unknown and no risk factors are identified in most patients. A small percentage (five to ten percent) of cases run in families. The average age of onset is between fifty to sixty years of age.
• How is it diagnosed?
A careful history and physical examination by a neurologist, along with some blood tests and x-rays to rule out other (rare) causes, will suffice in making the diagnosis.
• What are the pitfalls of diagnosis?
In cases where patients have only voice/speech problems, they may be sent to an otolaryngologist. Early stages of ALS can easily be missed by an otolaryngologist who does not have experience in treating ALS patients. It may be misdiagnosed as spasmodic dysphonia if vocal spasms predominate, or vocal fold weakness (paresis) if the flaccid symptoms predominate. Typically within weeks to months the disease progresses to involve the arms and legs, and the diagnosis becomes more obvious. A consultation with a neurologist results in the correct diagnosis being made.
• How is it treated and what is the prognosis?
There is no cure for ALS, only supportive care. The disease is variable, but fifty percent of patients die from respiratory failure within three years of onset of symptoms. With some slowly progressive forms of the disease, patients may live for ten years. In these select cases, an otolaryngologist may perform procedures to relieve vocal spasms (Botox injections into the vocal folds), or correct glottic incompetence with medialization laryngoplasty.

6. Myasthenia Gravis (MG)
• What are the symptoms?
The most common symptoms of MG involve the eyes, specifically double vision and drooping eyelids. The symptoms usually arise after repetitive tasks (due to muscle fatigue), and improve with short periods of rest.
Voice and speech-related symptoms include hoarseness, vocal fatigue, difficulty with controlling the pitch of the voice, hypernasal voice, mildly slurred speech, and monotone voice. Swallowing difficulty and weakness of the jaw muscles are also commonly encountered.
• What is the cause of the disease and who is at risk?
MG is an autoimmune disease affecting the nerve-muscle interface. The disease is more common in women (2:1), and presents between the ages of thirty to forty in females, and sixty to seventy years of age in males. No known risk factors exist.
• How is it diagnosed?
MG is diagnosed a number of ways, but the most reliable method is a blood test that detects the abnormal antibodies to the nerve-muscle receptor. Some investigators have pointed out that these antibodies may be undetectable in cases of isolated laryngeal MG. Another method is the so-called “tensilon test,” where the muscle in question is fatigued by a repetitive task, and edrophonium (tensilon) is given which dramatically but temporarily reverses the fatigue. Other tests look at the muscle electrical firing (EMG) during repetitive tasks and the muscle response to tensilon.
• What are the pitfalls of diagnosis?
The most common pitfall is failure to recognize the often subtle symptoms of MG. Because vocal fatigue is such a common component of many other voice disturbances, an otolaryngologist may misdiagnose the patient’s condition as vocal fold bowing, paresis of the vocal folds, or voice over-use/abuse. Evaluation by a neurologist should be obtained in unclear or unusual cases of vocal fold weakness/fatigue to rule out MG.
• How is it treated and what is the prognosis?
MG is treated with medications referred to as anticholinesterases
7. Multiple Sclerosis

- What are the symptoms?
  MS is a chronic neurological disease that is characterized by episodes of dysfunction of the nervous system that remit and recur over several decades. Commonly, long periods of normal function occur between these episodes.
  Voice symptoms may include hoarseness and poor control of volume and pitch. Speech problems are probably more common and have been characterized as “scanning speech,” where each syllable is produced slowly and hesitantly with a pause after every syllable.
  Other important symptoms include dizziness (vertigo) and altered vision.

- What is the cause of the disease and who is at risk?
  The cause of MS is unknown, but it is thought to be viral in origin. The process involves loss of the protective sheath around nerves in the brain/brainstem. The disease is more common in females, those of higher socioeconomic groups, and those living at greater latitudes (more rare at the equator). Onset of the disease is most frequent in young adulthood.

- How is it diagnosed?
  Diagnosis of MS may take years and is dependent on multiple signs and symptoms, with remissions and exacerbations of the disease. CAT or MRI scan may show the characteristic scar changes in the brain, and fluid from a spinal tap may also help in the diagnosis.

- What are the pitfalls of diagnosis?
  Because of the long latent periods, periods where disease is not present, and the waxing and waning of symptoms, MS is easily missed. Repeated examinations and clinical suspicion by a neurologist will help cinch the diagnosis.

- How is it treated and what is the prognosis?
  There is no cure for MS at this time. Care is directed towards controlling symptoms, and speech therapy plays an important role in improving quality of life in patients. Medical therapy includes corticosteroids for acute exacerbations of MS symptoms. For chronic treatment of MS, interferon and chemotherapeutics such as Novantrone have been used to slow the progression of the disease.

Areas of controversy/future directions

Myasthenia gravis is considered by most laryngologists to be a distinctly rare cause of isolated hoarseness. Some laryngologists have described “isolated” MG of the larynx which can cause hoarseness, vocal fatigue and reduced loudness, difficulty with pitch, and lack of vocal clarity—without any of the other eye, neck, jaw, or facial muscular weakness. These cases of “isolated” MG were mostly sero-negative, meaning the blood test for MG was negative. Many of the patients had improvement in their voice symptoms on MG medications, and subjective changes in their EMG testing that are somewhat compelling. It remains to be seen whether MG is a more common cause of isolated voice problems.

Patients with Parkinson's disease felt some hope in the late 1990s that a procedure had been developed to cure their vocal difficulties, but this has not materialized to any significant degree. Injections of collagen (autologous or bovine) do result in improved vocal fold contact, but most laryngologists believe this results in little or no benefit in the patient's voice. The primary problem with Parkinson's patients' voices is more due to the hypokinetic nature of their disease (reduced effort, reduced airflow) than to any loss of muscular tissue in the vocal folds. This probably explains the disappointing results with collagen injection and medialization laryngoplasty.

Traditionally, many progressive degenerative neurological diseases have not been referred to laryngologist/voice specialists, because it was thought not much could be done. In reality, as these patients' life expectancies and prognoses are improved, quality of life issues such as voice and communication become more important problems to the patient. Many excellent minimally invasive treatments currently exist to treat both vocal fold spasms (Botox) and vocal fold weakness (injection laryngoplasty/medialization laryngoplasty). In addition, in many neurologic diseases, speech-language pathologists and singing voice specialists can significantly improve the function of the speech, voice, and/or swallowing functions, resulting in enhanced quality of life.
SUGGESTED READINGS


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