

Spasmodic Dysphonia

Ken W. Altman MD, PhD

What is Spasmodic Dysphonia?

Spasmodic dysphonia (SD) is a neurologic disorder that is characterized by uncontrolled contractions of laryngeal muscles, manifesting as a "strained-strangled" or sometimes breathy voice. It is relatively rare, accounting for only about one to two percent of patients in a busy laryngology or voice clinic. SD is categorized as a laryngeal (focal) "dystonia." In contrast to generalized dystonias (movement disorders), focal dystonias are localized but may be exacerbated while performing a specific task. Examples of other focal dystonias include:

- Writer's cramp (occurs only while writing)
- Blepharospasm (eyelid twitching; increases with speaking)
- Oral mandibular dystonia (jaw-opening spasms that occur during speech, but not while chewing)
- Torticollis (turning the head and stiffening of the neck)

SD is not "spastic" dysphonia, because there are spasmodic electrical bursts in the larynx. Also, spasticity or rigidity (such as the result of a spinal cord injury) has not been demonstrated in patients with SD.

There are two manifestations of SD. Adductor type of spasmodic dysphonia (ADSD) accounts for sixty to ninety percent of patients, and is characterized by a tight closure of the larynx at the wrong time forcing the voice to stop. Abductor spasmodic dysphonia (ABSD) often manifests with a breathy voice. The spasms open the larynx at the wrong time rather than closing it.

What Causes Spasmodic Dysphonia?

We do not know how SD develops. It was originally described by Traube in 1871 as a form of "nervous hoarseness," and later referred to as "laryngeal stuttering." It is currently believed to be due to a dysfunction in the basal ganglia of the brain. Altered levels of neurotransmitters have been detected in autopsies of patients with other dystonias. There is also the theory that there is a central disinhibition of laryngeal responses to sensory input in patients with ADSD. Since SD is considered a focal neurologic dystonia and is usually confined to the larynx, problems with swallowing or other movements may suggest another neurologic diagnosis. At one time SD was thought to be due to a psychiatric disorder. Although stress can aggravate the symptoms of SD, the dystonias are considered neurological movement disorders.

How is Spasmodic Dysphonia Diagnosed?

SD presents in adulthood and affects two times as many females as males. Patients may first notice symptoms during a period of increased stress, following an upper respiratory infection, or during a speech performance. The voice usually sounds "strained-strangled," similar to the sound of someone being "choked." It doesn't hurt to talk, and it is usually worse when speaking loudly. Singing, crying, laughing and whispering often sound normal.

The larynx appears anatomically normal on examination by the otolaryngologist, but there may be subtle findings on the exam. Laryngeal electromyography (EMG) generally has a normal pattern of muscle activation with an occasional intrusion of spasmodic bursts, but there may also be prolonged spasms at a voice break. Since the laryngeal and EMG findings are not consistent among all patients, SD is a diagnosis by the otolaryngologist based on the clinical findings discussed above.

Characteristics of Adductor Spasmodic Dysphonia (ADSD)

ADSD is characterized by spasmodic vocal folds closings. Flexible fiberoptic visualization of the larynx may show intermittent rapid shortening and squeezing, resulting in a quick glottic closure (stop) that shuts the glottis, temporarily interrupting airflow.

Characteristics of Abductor Spasmodic Dysphonia (ABSD)

ABSD is characterized by wide-ranging abduction (opening) movements for voiceless consonants that are prolonged and interfere with the following vowel. Since these patients have difficulty with voice onset following voiceless sounds, there are prolonged voiceless consonants, that is, /h/, /s/, /f/, /p/, /t/, and /k/. There may also be pitch or voice breaks during vowels, with uncontrolled rises in fundamental frequency during speech, or especially a breathy voice quality.

There is no evidence that the two manifestations of SD represent different diseases. The two forms result from relative predominance of the main laryngeal muscle groups affected by the spasms.

Problems That May Make the Diagnosis More Difficult

- If patients also have a constant voice strain and strangle, then they may have a superposed muscle tension dysphonia.
- If patients report they have intervals when they are completely symptom-free, then they may have a psychogenic dysphonia.
- Patients may also develop compensatory speech patterns to improve their ability to communicate: whispering (less effort and greater control), speaking on inhalation (overcomes hyperadduction in ADSD, and produces voice more easily in ABSD).
- Vocal tremor, which may be related to benign essential tremor, is also focal and task-specific and may be confused with SD. However, when producing a prolonged vowel, there is regularity of the glottal stops or frequency and amplitude variation.

The Role of Botulinum Toxin Injection

Botulinum toxin is produced by the bacteria *Clostridium botulinum*. While it is a dangerous toxin in the natural environment, it has been shown to be safe and effective in treating SD when injected into the larynx by a trained professional. The medical use of botulinum toxin was introduced by Scott (1980) for the treatment of strabismus (eye muscle imbalance), and later used for blepharospasm (eye muscle spasm, Scott 1985).

During a botulinum toxin injection for SD there is a small amount of discomfort. Many clinicians use EMG to confirm appropriate needle placement. A small-gauge needle is used to confirm placement in the target muscle and to inject the botulinum toxin.

Effects may be noticed as early as six to 12 hours, but usually by one to two days and may continue for up to seven days due to diffusion of the toxin. Some patients may have difficulty swallowing liquids during this time (three to five days), and breathy speech may last from seven to 20 days. Most patients enjoy up to a two to three month period of improved speech without breathiness or difficulty swallowing. Since symptoms return due to new motor unit sprouting and reinnervation, injections are generally required every two to four months.

Other Treatment Options

- Voice therapy alone may be effective for patients mild forms of SD. Voice therapy also may provide support for patients unlikely to improve on other therapies, and may prolong the duration of effectiveness for those treated with botulinum toxin.
- Medical therapy may occasionally be effective for patients with SD. Propranolol (beta-blocker) "blunts" the effects of a voice tremor. Anti-cholinergics such as trihexyphenidyl (Artane) or benzotropine mesylate (Cogentin) may benefit severe dystonia. Muscle relaxants such as baclofen have some effect in a few patients. Central nervous system depressants such as diazepam (for example, Valium and Valrelease) or alprazolam (Xanax) reduce stress and its manifestations. However, these medications usually have discouraging results in patients with SD and they have potential side effects. Other treatments proposed for SD include hypnosis, acupuncture and electrical stimulation of the recurrent laryngeal nerve, none of which show definitive scientific proof of efficacy at this time.
- Surgery of the recurrent laryngeal nerve (RLN) includes section and avulsion (cutting the nerve and scraping the ends). The success is variable, however reinnervation occurs on the avulsed side and/or SD still affects the non-operated side. This operation is irreversible, and may leave patients with either dyspnea (difficulty breathing) or with a breathy voice.
- Other surgical options that have been proposed include selective thyroplasty, partial thyroarytenoid myectomy and anterior commissure retrusion, all reported to have limited or variable success.
- Support groups are especially helpful in patients with SD since people with a speech motor disorder may tend to have a compromised self-image and defer social engagements due to their difficulty with speech communication.

Conclusion

Spasmodic dysphonia is a focal neurologic dystonia occurring in the larynx. ADSD type occurs in 60 to 90 percent of patients, and manifests as a "strained/strangled" voice. ABSD type occurs less often, and manifests as a "breathy" voice. Botulinum toxin injection into the laryngeal muscles is the typical treatment.

Related Information

- National Spasmodic Dysphonia Association 1-800-795-NSDA
- Dystonia Dialogue: <http://www.dystonia-foundation.org/>
- Baylor College of Medicine - Adductor Spasmodic Dysphonia: <http://www.bcm.edu/oto/grand/71391.html>

Author

Ken W. Altman MD, PhD