

The Voice and Swallowing Institute

Spasmodic Dysphonia (SD)

Spasmodic dysphonia (SD) is a type of dystonia, a class of disorders of central motor processing (the part of the brain that controls movement) that cause involuntary movements in the affected body part. Dystonias can affect the entire body (generalized dystonia) or just one part (focal or segmental dystonia). SD is a dystonia that is limited to the larynx. Examples of other dystonias include blepharospasm (involuntary eye blinking) and torticollis (wry neck). Among neurologists, SD is known as focal laryngeal dystonia. Spastic dysphonia is an older term that is synonymous with spasmodic dysphonia. There are two types of SD. Adductor spasmodic dysphonia (AdSD), so named because it affects the muscles that bring together (adduct) the vocal folds during speech, is the more common of the two (85-90% of cases). Abductor spasmodic dysphonia (AbSD) affects the opposite muscle group, which moves apart (abducts) the vocal folds during speech. These involuntary movements cause the symptoms of SD, which are discussed below. SD usually occurs by itself, and is very rarely associated with other, more serious neurologic diseases.

People affected: SD usually affects adults, with typical onset of symptoms in the 30s. Onset earlier or later in life is not unusual, however. Women are more often affected than men. All ethnic groups are affected. SD does not seem to be hereditary. There is a greater incidence of SD in Caucasians who are of the Ashkenazi Jewish ethnic background. The incidence of general dystonias in the world population appears to be increased in Caucasians and a lower in Asians and persons of African descent. It is unclear if the incidence of SD shows the same pattern.

Causes: There do not appear to be any behaviors or environmental factors that increase the chance of contracting SD. Researchers believe that SD stems from abnormal brain cell activity in a part of the brain called the basal ganglia. The cause of this is unknown. SD, like all focal dystonias, is a disorder of unknown cause. Affected persons have reported the onset of symptoms following trauma to the head, common upper respiratory infections, routine intubation for general anesthesia and dental work. However, no clear relationship between any of these conditions and SD exists.

Symptoms: Involuntary, abnormal motion of the vocal folds (spasms) causes the characteristic voice changes of SD. In AdSD, the vocal folds come together with too much force and at inappropriate times, producing strained, strangled breaks in connected speech. In AbSD, the vocal folds spread apart inappropriately during speech, causing breathy or soundless breaks. In both cases, voice breaks, or spasms in the voice occur irregularly. The voice will usually "break" or be interrupted on specific sounds and parts of the word. This occurs typically during voiceless to voiced transitions during speech (i.e. "T" and "P" sounds followed by a vowel). For example, when an AbSD patient says the sentence "Pay Paul a penny" there will usually be a breathy,

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voice break immediately after each "P".

The severity of the symptoms usually varies from day to day, and even over the course of a single day. It is possible for the voice to be normal sometimes, and affected at other times. It is clear that anxiety or fatigue causes symptoms to be more noticeable.

Speaking to strangers, public speaking, or speaking in unaccustomed situations often makes symptoms worse, probably because all of these situations increase anxiety.

Most people with SD report that using the telephone is especially difficult. Conversely, symptoms are usually mildest in familiar situations. Anything that functions as a sedative, including a glass of wine or beer, can improve symptoms.

Most people gradually become aware that their speech is requiring more conscious effort. Over weeks to months, the characteristic voice breaks become more noticeable and pronounced. The severity of the voice disturbance may vary considerably in the initial stage of the disorder. Over time, it becomes more consistent. There is no evidence that the disorder worsens over the course of a person's life, nor, unfortunately, is there evidence that it improves or resolves. Patients with SD almost never lose completely the ability to communicate.

Diagnosis: There is no specific test to diagnose SD. Diagnosis is based on the patient's description of the symptoms, the sound of the voice, and the physician's observation of the vocal folds during speech. For this reason, the physician will likely take a detailed history that touches on the features of the disorder mentioned above. Because SD is a central nervous system disorder, the physical exam should include a careful neurologic examination. Some otolaryngologists will ask that a person with suspected SD see a neurologist for this.

The most important part of the examination is observation of the vocal folds during speech using a flexible fiberoptic laryngoscope inserted through the nose. The vocal folds will demonstrate the abnormal behavior typical of SD as the patient speaks. This part of the examination serves to distinguish SD from conditions that can cause similar voice changes. In addition to a careful history and physical examination, some otolaryngologists may use acoustic and aerodynamic measurements, and/or electromyography to help diagnose unusual cases. It is important to understand that there is no specific finding on any test that identifies SD. Although these tests may be helpful, the diagnosis of SD is ultimately a matter of expert opinion, formed from information gathered from the history, physical examination, and, most importantly, the otolaryngologist's assessment of the nature of the sound of speech.

Errors in Diagnosis: Because cases of SD can appear very different from one another, an inexperienced examiner can mistake SD for a number of similar conditions that have very different treatments. Perhaps the most unfortunate mistake is to ascribe symptoms to psychiatric problems. The symptoms of SD are so variable and some compensatory

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behaviors so unusual that uninformed observers may conclude that they are the result of a psychological disturbance. Fortunately, most specialists (otolaryngologists and neurologists) are now familiar enough with SD that this error is not often made. SD is beyond question a physical disorder of the motor processing centers of the brain, and not a psychological problem. Although psychiatric counseling sometimes is suggested in managing the anxiety that often makes symptoms worse, it is not a useful treatment for SD. SD may be mistaken for essential vocal tremor. Severe cases of vocal tremor may cause voice breaks and stops very similar to those of AdSD. In contrast to SD, however, these usually occur with rhythmic regularity, and persist across all types of voicing, including speech, sustained vowels and singing. Tremor may affect the muscles of the pharynx and the muscles on the outside of the larynx (the strap muscles or extrinsic laryngeal muscles), whereas SD is usually limited to the muscles of the vocal folds.

Tremor is primarily treated with medications, an approach that has little benefit in SD. In approximately one-third of cases of SD, there is a something known as dystonic tremor. This is a combination of spasmodic dysphonia with tremor, which can make the diagnosis confusing. AdSD can occasionally be mistaken for **muscle tension dysphonia**. The squeezing together (hyperadduction) of the vocal folds that is the hallmark of muscle tension dysphonia is very similar to that of SD. However, SD is, as its name says, spasmodic, meaning that squeezing takes place irregularly, whereas squeezing is generally sustained in muscle tension dysphonia. Nevertheless, the distinction can be unclear, and in rare cases, a physician may elect to proceed with treatment for one disorder or the other, and use the outcome of treatment to arrive at a definite diagnosis. The primary treatment for muscle tension dysphonia is voice therapy, which is only of limited benefit to patients with SD.

SD can be present simultaneously with other voice disorders, making the diagnosis difficult. SD is often seen with tremor as mentioned above. When this occurs, the patient's speech has both spasmodic speech breaks during speech (SD) and a "tremorous" wobble or "shake" to the voice. Muscle tension dysphonia and SD can often occur together as well. Most believe the muscle tension dysphonia is the body's attempt to compensate for the SD. The concurrent presence of these two disorders can make the diagnosis of SD difficult. The primary disorder will usually become evident after an otolaryngologist takes a careful history, listens to the voice and performs a physical examination. He/She may suggest an evaluation by a speech language pathologist or a brief trial of voice therapy to aid in the determination of the difference between SD and muscle tension dysphonia. Patients with primarily SD will not significantly improved with voice therapy and those with muscle tension dysphonia will benefit from voice therapy.

Treatment: There is no cure for SD. Available treatment is intended only to improve symptoms, and happily, that is almost always possible. It is important to understand that

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treatment does not affect the course of the disorder. In other words, if a person elects not to be treated, the SD will not become worse.

Voice Therapy. Voice therapy, by itself, has not been useful in controlling the symptoms of SD. Voice therapy can help control side effects of treatment discussed below, and can help a person manage the anxiety that often makes the symptoms of SD worse. Voice therapy may be helpful following **botulinum toxin treatment** (see below) by helping the person to: (1) eliminate the body's poor compensation behaviors; (2) adjust to the changes in the voice from treatment and; (3) maximize the beneficial effects of the treatment.

Psychological/Psychiatric treatment. SD is an organic disorder of the central nervous system. As a result, psychological/psychiatric treatment is not an effective primary treatment. However, any chronic condition can be emotionally and psychologically draining. Because SD affects the voice, it is especially so. A mental health professional may help affected people better handle the stresses associated with this disorder.

Medical treatment. Many medications that act on the central nervous system may be beneficial in dystonia. Unfortunately, at doses that relieve symptoms, there are often significant side effects like sedation and memory loss. Direct treatment of the involved muscles has generally proven to be of greater benefit. In most cases of SD, oral medication is not used at all, or is used only as an auxiliary therapy.

Botulinum toxin. Laryngeal injections of botulinum toxin are the main therapy for SD. Botulinum toxin is a naturally occurring substance that weakens muscle by blocking the release of acetylcholine from nerve endings. Acetylcholine is a substance that triggers muscle contraction. The effects of botulinum toxin are temporary, lasting about three months, and dose-dependent, so that the muscle weakness is proportional to the amount of toxin used. Research has found that there is a "plateau effect" at higher doses of botulinum toxin; however, at the typical low doses used for the treatment of SD, there is a consistent dosedependent response. Of the eight types of botulinum toxin that exist, two are available for use in humans - botulinum toxin type A (Botox®, Allergan Inc., Irvine, California, and Dysport®, Ipsen Ltd., United Kingdom) and botulinum toxin type B (Myobloc®, Elan Pharmaceuticals, Dublin, Ireland). Botox® is the formulation most often used in the United States.

Botulinum toxin has been used in humans since 1979, and in SD since 1984. In that time, it has been shown to be safe and effective in the treatment of SD, and is recognized as such by the American Academy of Otolaryngology - Head & Neck Surgery. Botulinum toxin is not approved by the United States Food and Drug Administration for use in SD, and none of the companies that produce it have plans to pursue such approval, probably because the relatively small amount used for SD makes

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this process financially unrewarding. Because botulinum toxin use in SD is not FDA-approved, the manufacturers are not permitted (in the U.S.) to discuss information related to this use. Nevertheless, general information is available from each company (see below). The principle behind botulinum toxin treatment of SD is to weaken the muscles that are hyperactive. In the case of AdSD, these are muscles that bring the vocal folds together, and in AbSD, muscles that bring the vocal folds apart. Botulinum toxin is usually injected through the skin of the neck into the appropriate spots with the aid of electromyography (EMG). (LEMG link) This is an office procedure. Afterwards, the patient may usually go on with the normal activities of the day. The discomfort associated with the injection commonly disappears after a day or two.

In AdSD, because the muscles that bring the vocal folds together are initially "over-weakened," injection is normally followed by a period of breathy, whispery voice and sometimes coughing when drinking liquids. This may last for up to two weeks. Most otolaryngologists aim to adjust the botulinum toxin dose to limit the "breathy" period to one week. In AbSD, the situation is more complicated, as inadequate parting of the vocal folds might result in difficulty breathing. For this reason, AbSD injections are often staggered, with one vocal fold injected one or two weeks after the first. Sometimes, a treating physician may prefer to treat just one side per three-month "cycle" to minimize these effects. Often, the dose of botulinum toxin can be adjusted to minimize unwanted effects. In the case of AdSD, for instance, decreasing the dose can usually shorten the duration of the breathiness, but the overall length of benefit may be decreased. Obviously, each person prefers to be injected as infrequently as possible, but each has a different tolerance for the breathy voice period following the injection. People to whom voice is crucial, like lawyers or schoolteachers, may opt for smaller doses at more frequent intervals.