Dr. Steven Bielamowicz from George Washington University in Washington, DC, kicked off the day’s presentations with an Overview of Spasmodic Dysphonia.

Dr. B. told attendees SD is neurological. It doesn’t appear to be a laryngeal disorder. It is also a movement disorder in the same family as writer’s cramp.

There is no known cause of SD, and patients should know that there is nothing they did to bring this on themselves. Also, SD is not an emotional illness. Before the 1970s, it was thought to be psychological in nature. Not all patients can point to a traumatic event in their lives that would have brought on the condition.

Although there is a likely genetic basis for SD, it’s not directly inheritable, so it’s not a given that a patient’s children will get it.

No specific diagnostic test exists for SD. Doctors know you have the condition by how you sound. In adductor SD, the more common type, patients have a strained or strangled voice. In abductor SD, which accounts for 10% of cases, there are prolonged breathy breaks after the voiceless consonants (h, k, p, t).

Speech therapists can elicit the symptoms of either type by asking patients to read out sentences with particular combinations of vowels that follow consonants. The consonants can either be voiced or unvoiced:

Sentence 1 (causes problems for AD): ‘We mow our lawn all year’

Sentence 2 (causes problems for AB): ‘Keep Tom at the party’

One of the most frustrating things for SD patients is not knowing how their voice will sound from day to day, or even from moment to moment.

Dr. B. said this is part of the nature of the condition. While SD isn’t caused by anxiety, feeling anxious can make it worse. Anxiety can make any type of motor disorder worse. An example: At the 1984 Olympics in Atlanta, Mohammed Ali was in the early stages of Parkinson’s Disease. He didn’t have much of a clinical tremor at the time, but when he went to light the torch in the stadium, the anxiety of being on such a large world stage caused him to shake more severely.

Another question patients often ask, according to Dr. B., is, “Will my SD ever go away?” The answer is, if you’ve been correctly diagnosed, then, no. Dr. B. says in all his patients in 20 years, he can only think of two who have had a long-term recovery from SD. These are patients who had long-term responses to Botox treatments.

The second speaker, Dr. Christopher Chang of Fauquier Ear, Nose & Throat Consultants in Warrenton, VA, addressed the Use of Botulinum Toxin for the Treatment of Spasmodic Dysphonia.

One common question from patients that Dr. Chang often fields is, “Why did one doctor do my Botox injection this way, and another doctor did it a different way?” The answer is that there is no right way. Doctors perform the injections in the way that works best for them. Sometimes there is assistance, sometimes not.

Here’s one common way the injections work for patients with adductor SD: The needle is injected through the front of the neck and straight into the muscle inside. The doctor first injects a numbing agent into the airway. It helps the patient remain still during the shot.

The sound from the EMG (electromyography) changes when the needle reaches the muscle. That’s when the Botox is injected. If you receive bilateral injections, the doctor may
keep the needle inside and move it to the other side for the second injection.

Another method uses a scope when someone has an unusually thick neck. An assistant holds the scope steady while the doctor looks at a monitor to perform the injection.

Botox shots are also used for abductor patients. Dr. Chang says he has only about a 50% success rate with these SD patients. The muscles that are targeted are behind cartilage and are flatter. That’s why it’s more difficult to hit the correct spot. Injections for abductor can be done through the front or through the side. The side shots can be uncomfortable for the patient because the larynx has to be physically moved aside.

Should you get a larger or smaller dose? People who use their voices professionally, such as lawyers, may prefer a smaller dose so the soft voice period afterward is shorter. People who live in rural areas and may have to travel great distances to get their injections might prefer larger doses so they last longer.

Another variant besides the concentration of Botox is the amount of dilutant used to make up the solution. Most doctors use 4 ccs. Dr. Chang uses 6 ccs, and a survey found doctors using anywhere from 2 to 10 ccs.

For patients who receive a larger dose of Botox, distributed injections are an option. The doctor inserts the needle, injects some of the solution, then moves the needle a little further and injects again. Effectively, there are two injections with a single needle.

Another treatment for adductor SD is surgery. Dr. Young Kim with Johns Hopkins Hospital in Baltimore spoke about SLAD-R (Selective Laryngeal Adductor Denervation-Reinnervation) surgery.

Who chooses surgery for SD? Some patients want the surgery because Botox injections no longer work. For others, it’s because the Botox cycle can become annoying.

SLAD-R was pioneered by Dr. Gerald Berke. It’s performed under general anesthesia. An incision is made in the mid-portion of the neck, about 4-6 cms (1.5 to 2.3 inches). A small window is created to expose the nerves and muscles. Then the surgeon cuts the nerves to the muscles on both sides of the larynx. Next, the surgeon connects an unaffected nerve to the ends of the nerves severed in the muscles. The surgery takes from three to six hours.

Data from Dr. Berke shows 90% of SLAD-R surgery patients report an improvement in their voices. The patients are pleased with the smooth articulation of their voice. There is a 10% failure rate.

One downside: There is a prolonged soft voice period of anywhere from six months to slightly more than a year.

Dr. Berke tried some unilateral SLAD-R surgeries, but they weren’t successful. The surgery isn’t for patients with abductor SD.

One of the most common questions from people with SD is, “What causes this condition?” Dr. Mark Hallett with the National Institute of Neurological Disorders and Stroke (NINDS) at the National Institutes of Health (NIH) in Bethesda, addressed that question in his session, *Looking at the Big Picture: Possible Causes of Dystonia.* (SD is a type of dystonia.)

A recent committee has developed a revised definition of dystonia: A movement disorder characterized by sustained or intermittent muscle contractions causing abnormal and often repetitive movements, postures or both. Dystonic movements are often patterned, twisting and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with muscle activation.

This definition is relevant to SD because there is an abnormal contraction of the muscles that deal with the vocal cords and it’s activated by...
attempts at talking. One part isn’t relevant to SD: There is no twisting in SD.

Dystonia is multi-factorial, that is, there are multiple causes. Genetic background is relevant.

Gene modifiers (these are able to alter expression of a gene) affect cell biology. The cell biology leads to regional anatomical abnormalities. This relates to disorders in particular regions on the brain. The brain operates in networks, and those networks lead to physiological problems.

Where are we in this chain when it comes to dystonia?

More and more genes have been identified that produce dystonia, the DYT genes. We’re up to DYT-25, with the three most recent genes identified in just the last year. DYT-24 may be of interest for SD. People who have it have cranio-cervical (head and neck) dystonia which includes laryngeal dystonia.

The next step is knowing what the genes do. For example: Researchers have been working for years to figure out what DYT-1 does and how it affects certain abnormalities.

This is still a long way from understanding how you get SD or another type of dystonia. And this is about as far as this research has progressed at this moment.

Studies have been performed to identify the regions of the brain that may be abnormal in SD patients. One study by Dr. Christy Ludlow identified abnormalities in the internal capsule of the brain which sends information from the cerebral cortex to the brainstem that controls the speaking muscles.

Abnormalities have also been found in SD patients in the basal ganglia and cerebellum. The basal ganglia is important to dystonia because it’s a key network in movement.

This is the way the brain makes a selective movement: The brain issues a specific motor command for the movement, and at the same time it issues commands to inhibit movements you don’t want.

It turns out the inhibition is a bit of a problem for patients with dystonia. If you have a good motor command but the inhibition is lacking or you get too much, you get failure of the movement you’re seeking. This is controlled in the basal ganglia.

How can there be a loss of inhibition? Something called GABA (gamma-amino butyric acid) is responsible for this. A paper will soon be published on GABA. GABA abnormalities seem to be relevant to the production of overflow movements.

Just as researchers are studying all types of dystonia, there has been an explosion of research into SD. Dr. Christy Ludlow, with James Madison University in Harrisonburg, VA, gave the Research Update on Spasmodic Dysphonia.

In 2005, there were five grants for SD research supported at the National Institutes of Health. Now, in 2013, we have 13 grants. The emphasis is on the brain and neurological system with SD, with seven grants in that area. Six other grants are looking into other issues, including a measurement of the impact SD has on patients’ lives, treatment using sodium oxybate and an attempt to develop a test for SD.

Dr. Ludlow said in 2009, a study was started to develop a test for SD. The objective was to develop a method of identifying patients with adductor SD, abductor SD, muscle tension dysphonia and tremor.

A gold standard for identifying what is and what isn’t SD was needed. A panel of four judges looked at videos and weighed in on whether the people in the videos had SD or not. There wasn’t enough agreement among the four experts.
A statistician suggested a solution called the Delphi method. A list of potential attributes for abductor, adductor, tremor and MTD were made. Experts in various fields related to SD examined the lists. The result: There were two top attributes for each condition that pretty much all of the experts agreed on. This project should finish in about a year.

Why is this work to identify a test for SD important? We can’t do clinical trials in SD without a measurement tool that would allow comparisons of results of various treatments.

What are some of the other outcomes of the research to develop a test for SD? Dr. Ludlow hopes it improves accuracy of diagnosis so patients can get treatment faster. This will also allow researchers to better study brain function in SD. Eventually, Dr. Ludlow says it may be possible to modify the brain in people with SD and even identify ways to avoid its onset.

An afternoon session at the symposium looked at legal issues, particularly when it comes to employment. Two speakers addressed this issue.

The first was Teresa Goddard with the Job Accommodation Network (JAN). JAN is a service offered by the U.S. Department of Labor. It’s the leading source of free, expert, and confidential guidance on workplace accommodations and disability employment issues. Teresa fields questions from employers and employees about the workplace under the Americans with Disabilities Act (ADA).

One of the basic strategies for dealing with a vocal problem in the workplace is to improve the signal to noise ratio: making things you don’t want to hear softer so people can hear your voice. Example: Foam that adheres to a wall can lower the ambient noise in a room. Cubicle environments can be difficult. Moving to a quieter location at work might help. Example: A noisy place is near a copying machine.

Assistive technology can also be a solution. A personal microphone device can help as can equipment to amplify your voice on the telephone. Teresa says JAN has a list of assistive technology on its website (http://askjan.org/), but she advises that you check with each equipment vendor because they add and delete features all the time.

What’s the best way to procure accommodations at work? Teresa advises that using the ADA to “hit an employer over the head” might not work so well. But if you use it as a tool to show there are accommodations that are inexpensive and will help you be more productive at work, you’re more likely to get cooperation from your employer.

Although the ADA does not require you to put a request for an accommodation in writing, Teresa advises that you do so. It’s been the experience at JAN that writing a letter helps. It establishes a record and shows the seriousness of your request.

How does seeking accommodation work? Here’s one example. A service technician had to troubleshoot over the phone. He was having problems speaking. The employer restructured his job on a temporary basis. He was relieved of troubleshooting over the phone while he underwent treatment. He was able to keep his job while exploring treatment options. His employer said this accommodation cost nothing to do, and the benefit was retaining an employee.

Does everyone with SD fall under the ADA? Teresa said she couldn’t say for sure. Here’s why: There’s a functional definition of disability: impairment which substantially limits a major life activity. Speaking is on the list of major life activities. However, there are no conditions that are always covered.

The second speaker in the session on employment was Judge Maryellen Halversen, an appeals judge with the Social Security Administration (SSA).
Most people might think about SSA in regard to retirement benefits. But the agency also administers the federal government programs for disability coverage. Last year, the SSA handled three million disability applications.

Disability benefits come under two different areas: Title 2 and Title 16.

To qualify for Title 2, first you need to have insured status: You must have 10 quarters of FICA coverage through working in the last 20 years. The SSA will help you figure that out, or you can calculate it on SSA's website (http://www.ssa.gov/).

Title 16 is different in that it’s designed for people who have very low resources and virtually no income at all.

Of all initial applications that are filed at the state level, about 33% of claims are allowed.

If your initial application is denied, you can file for a reconsideration. It gets looked at again, but only 11.7% of claims at the second level are successful.

If you are denied at that second level, you can file a request for review before an administrative law judge. The ALJs hold hearings at offices across the country. You can attend in person or through telephone or video conference.

You can call witnesses, present evidence and have representation at these hearings.

If you are turned down at the appeals level, you can request review by the appeals counsel (that’s what Judge Halversen does).

This is what is critical: How is disability defined by SSA? It means you can’t do any jobs existing in significant numbers in the national economy. It’s a high standard to meet. There are five steps:

1. Are you working? If you are, are you working full-time? If you are, then you do not qualify. If you are making more than $1,040 per month, you won’t qualify.

2. Is your condition severe? Judge Halversen guesses that SD would be considered a severe impairment, because severe is defined as anything that affects basic work activities. If you have any difficulty with speech, that’s a basic work activity.

3. Is your condition found in the list of disabling conditions? SD doesn’t have a particular listing. However, there are some conditions that may apply, such as No. 2.09, total inability to produce speech by any means that can be understood. There are also neurological listings, including dystonia.

4. Can you do the work you did previously? Are you able to do your job as you did, or as it’s generally performed in the national economy?

5. If you can’t do your past relevant work as actually performed, SSA will see if there are other jobs in the national economy you can perform. If you have a high school education, you’re considered pretty well educated, and there are probably a number of jobs you can do. There are extra considerations for people over the age of 55 who don’t have a high school education since manual labor jobs become more difficult as we age.