Essential tremor is a neurological disorder that presents with trembling of the voice as well as other body parts. When a patient shows signs of essential tremor along with tremulous voice, a feature of many movement disorders, the condition is known as Essential Vocal Tremor (EVT). It has been estimated that up to 40% of individuals diagnosed with essential tremor also present with EVT.

In EVT, there are alterations in the pitch and loudness of the individual’s voice because muscles in the larynx, pharynx, and the palate are fluctuating at a 4-8 Hz frequency. The manifestations of these fluctuations are most noticeable during sustained enunciation of vowels. Unfortunately, this causes patients to experience marked effort with their speech that worsens with stress and anxiety. This makes living with EVT difficult and sometimes embarrassing, and patients may find themselves avoiding school, work, and other social situations other people take for granted. Moreover, basic activities including writing, eating, and drinking can become difficult, significantly impacting an individual’s quality of life.

Management of EVT is difficult with current treatment options, as many patients do not experience substantial improvement. Deep Brain Stimulation (DBS) has been utilized for decades in the treatment of movement disorders like Parkinson’s disease and essential tremor, and has been proposed as a potential remedy for EVT. However, very few neurosurgeons have employed this technique specifically for the treatment of EVT. (Figure 1)

However, the physicians and researchers at Stanford University School of Medicine in California have been applying DBS in the treatment of patients with severe voice tremor, and for the first time are performing comprehensive vocal tremor evaluations, to maximize its effects.

Assessment and Treatment
EVT is currently assessed through the combined efforts of an otolaryngologist (ear, nose, and throat doctor) specializing in assessment and treatment of voice disorders, and a speech language pathologist, who helps the patient develop their phonation, intonation, fluency, pitch, and voice.

Currently, EVT is identified, quantified, and characterized by either transnasal or oral endoscopy. Endoscopy involves inserting a camera into the patient’s throat to visualize the vocal cords, and having them sustain a vowel sound, in order to observe any abnormal movements. Transnasal endoscopy, or endoscopy through the nose, is the preferred method, since the vocal cords are in a more natural position with this technique. A diagnosis of laryngeal tremor is made if the vocal cords exhibit rhythmic movements, present both during quiet breathing and during the pronunciation of a vowel. Patients typically also exhibit contractions of the mouth and throat muscles in an up-and-down direction.

In addition to endoscopic techniques, a voice evaluation is also performed with the aid of a speech language pathologist, which involves quantification of the abnormal rhythmic movements. (Figure 2) In particular, the rate and magnitude of these movements are used as benchmarks.
Treatment of EVT is initially managed with medications such as Propranolol and Primidone. If necessary, Botulinum toxin injections, also known as Botox™, are administered into the vocal cord muscles. However, this method will only be successful if the tremor is caused by the muscle that has been injected, and involvement of other untreated muscles will allow the condition to persist. Further treatment with the aid of a speech language pathologist has provided beneficial results for some patients, as they learn to adapt to their condition and master control over their vocal cords.

**Deep Brain Stimulation for EVT**

Unfortunately, the current treatments and interventions available for treating vocal tremors are limited and often not effective. Deep Brain Stimulation (DBS) has been hypothesized for many years to have potential to treat vocal tremor, but there have been few studies that demonstrate its effectiveness in a comprehensive and directed fashion. There are a handful of studies that demonstrate elimination or decrease in vocal tremor using DBS in the treatment of patients with essential hand tremor, Parkinson’s associated tremor, and dystonia. These studies indicated that vocal tremor did not improve as much as tremor in other parts of the body.

At Stanford, neurosurgeons work with the otolaryngologist and speech pathology team, using DBS to target a specific region of the thalamus known to be involved with vocal control. To the researcher’s knowledge, this marked the first time that a tailored approach was utilized in administering DBS treatment for vocal tremor. They determined stimulator placement during the operation, after careful evaluation with voice assessments in the operating room with a speech language pathologist.

**Target for DBS**

The specific region of the thalamus that was targeted for the DBS procedure is known as the ventralintermediate nucleus (Vim). The typical target for implantation of the DBS electrode for essential tremor is within the kinesthetic fields corresponding to the hand. The area of the Vim involved in head and neck movement, including the vocal cords lies more in the middle. Targeting this specific region is challenging, as it is very small and requires mapping with advanced imaging technology and intraoperative physiologic neuromonitoring for accurate placement. Keeping patients awake for the procedure allows for this neuromonitoring to confirm the reduction in tremor frequency and intensity with test stimulations. The desired effect in terms of reduction in vocal tremor is assessed by a speech language pathologist in the operating room to optimize and confirm the best position for final DBS electrode placement. (Figure 3)

**Conclusion**

Stanford University School of Medicine’s treatment of EVT utilizing awake, frameless DBS, guided by voice analysis has been successful. Patients’ vocal tremor were well-controlled, without any apparent negative effects. Further study is needed in the future to address tremor that occurs in multiple muscles, as they may require stimulation in additional sites in the brain. Larger clinical trials are also needed to establish this treatment as a standard of care. Finally, a multidisciplinary approach with rigorous pre-operative, intra-operative, and post-operative voice testing can be expanded to other voice disorders, including those related to Parkinson’s disease and dystonia.
Deep Brain Stimulation is a treatment option that has been underutilized for treating patients with EVT. Few studies have examined the results of DBS on vocal tremors, and none have tailored their interventions to target vocal tremors specifically. Stanford’s approach to specifically target the centrally-located Vim for EVT has yielded encouraging results. These findings will have tremendous value for these people, and will hopefully usher a new era of DBS-based treatments for people afflicted with vocal tremor.

For a complete reference list for this article, please contact the IETF office at 888.387.3667 or email info@essentialtremor.org.

Tomorrow’s Surgical Options
May be here sooner than we thought

By Catherine G. Kernie, MD; Pejman Ghanouni, MD; and Casey H. Halpern, MD

Essential tremor, the most common movement disorder, has a profound impact on people’s quality of life in both their home and workplace. Prescription medications, such as Propranolol and Primidone, can effectively treat tremor in some, however, up to 40% of people with essential tremor cannot tolerate or do not respond well to these options.

If medications fail to reduce tremor, there are surgical options available. However, not every individual who has ET is a good candidate for surgery. Research indicates that MRI guided high-intensity focused ultrasound (MRgFUS) may be used to treat patients with essential tremor who do not respond to the typical medications and who do not qualify for Deep Brain Stimulation or Thalamotomy. Although more research needs to be done, focused ultrasound will hopefully provide an alternative treatment option for people with essential tremor in the future.

MRgFUS may become an appealing surgical alternative because it is less invasive than other interventions, as no incision is required. In the case of essential tremor, the ventral intermediate nucleus of the thalamus (Figure 1) on one side

Figure 3. This section of the brain shows the location of motor receptors of the leg (open triangle), arm (open square), and face (open circle). The final placement of the DBS lead is represented by the thin blue line. Notice how the lead touches both the hand receptors and those of the face, thus effecting both hand and vocal tremor.