

Neurolaryngology

1. What am I doing?

I'm careful to examine all of them muscles innervated by the Xth cranial nerve, not just the vocal cords.

2. What I am doing no more?

I no longer perform electromyography. Although it is a valuable tool, it is a cumbersome examination, uncomfortable for the patient, difficult to interpret and careful endoscopic evaluation seems at least as precise.

3. What should I start to do?

Examine all of the upper airway, make use of topical anesthesia for a close detailed exam, observe slowly during both respiration and phonation.

In this lecture, the audience could expect to learn the following:

Visual diagnosis of laryngeal neurologic impairments is not only possible, but is perhaps the most accurate method for evaluating the neurologic status of the upper airway.

A complete endoscopic neurolaryngology examination begins in the mouth and nasopharynx. It includes movement of the pharynx and finally a detailed examination of the motion of the intrinsic laryngeal muscles. Principles of the neurolaryngology examination include:

1. each muscle has only a single action,
2. that action can be isolated,
3. that action can be elicited and visualized,
4. the patient always attempts to compensate and the examiner's goal is to remove compensation in order to better visualize the neurologic impairment.

Some of the concepts covered in this lecture will include endoscopic visualization of;

- paralysis
- paresis
- synkinesis
- fixation
- tremor
- spasm
- reinnervation

Extensive use of video recordings will support the precepts.

Principles of visual neuroanatomy

EMG

Although the EMG is thought to be the “gold standard” (if there even is one), visual examination offers a great deal - and might even prove to be a better gold standard.

We need a map.

The first step in visual neurolaryngology is to have a mental map of the anatomy in the examiner's mind. Understanding the treelike branching the nerve will allow us to work both forwards and backwards from clinical information we have. If we know where and how an injury took place along the path of the nerve, then we will know the findings and impairments that we should see on an endoscopic examination. Likewise we can work in the reverse direction. Based on the functional impairments that are seen on an endoscopic examination, we should be able to predict where along the path of the nerve the injury took or is taking place.

The larynx is supplied by the Xth cranial nerve, the vagus nerve. There are two important anatomic locations along the path of this nerve for the examiner to think about. The first location is where the 10th cranial nerve leaves the skull and heads towards the neck. It passes through a very narrow opening along with two other cranial nerves, the XIth and the XIIth cranial nerves. From a diagnostic perspective, this bottleneck then is a location where an injury to the nerve will likely affect not only the Xth cranial nerve but also the XIth and the XIIth cranial nerves. So in our examination, if there are problems with the tongue (XIIth cranial nerve) or with lifting the arm and shoulder up (XIth cranial nerve), in addition to problems in the larynx, we will know to focus our attention on this narrow opening at the base of the skull.

The second diagnostically important location is where the Xth cranial nerve descends in the neck, it gives off four branches to supply the throat and larynx. Knowing which of these branches are affected, will tell us how high the location of the injury is in the neck. If only single branch of the nerve is involved, we know the injury is taking place very near to the larynx. If multiple branches are involved, we know the injury is taking place further away from the larynx and closer to the brain.

Of course the most commonly understood anatomic asymmetry, the path of only the left recurrent laryngeal branch of the 10th cranial nerve descending into the chest and wrapping around the aorta and left bronchus while the right typically remains recurrent in the lower neck is one of the most known diagnostic asymmetries.

Regrowth

A second principle important in laryngology is that the Xth cranial nerve, when injured, almost always tends to regrow. Mild injuries and injuries close to an individual branch of the nerve will tend to recover with normal function. More severe injuries and we could say more proximal injuries, that is, injuries that are closer to the main trunk of the nerve, will result in regrowth with the crossing of nerve pathways to different branches. This will result in various degrees of mixed up functioning of the nerves and muscles of the larynx. Even in cases where there is a gap between the cut ends of the nerve, the cut neurons will often gradually grow through the intervening tissue and reconnect with the denervated muscles.

Bilateral symmetry

In many injuries of the larynx, we have a built in reference point. In the more frequent condition when one recurrent laryngeal nerve is injured, the other side continues to function normally and provides the examiner with a direct comparison of normal anatomy and normal function between the two sides. We can compare the structure on the left with the structure on the right. We can also compare the timing and degree of movement on one side with the other.

Observing muscles

Atrophy - TA, PCA

Fasciculation - TA, PCA

Oscillation - TA, CT

Tension - TA, CT

Range of motion - LCA, PCA (joint, scar)

Lengthening - CT (SLN, joint, scar)

Hyperactivation - LCA

Timing - inappropriate reinnervation

Compensation - unmasking

Larynx motion appears to be complex, at least on superficial examination and reference articles typically fail to discriminate among the various types of motion.

There are 5 types of mobility;

1. Opening - abduction - PCA
2. Closing - adduction - LCA (and for Maintaining closure - IA)
3. Tensioning (isometric) - TA
4. Lengthening - CT
5. Oscillation - passive flexibility

Definitions of movement

Paralysis

Loss of voluntary movement (motor)

Paresis

Partial loss of voluntary movement

Synkinesis

Simultaneous movement

Dyskinesis

Inappropriate movement (degree or timing)

Cranial nerve X examination

Upper ganglion branch - supplies palatal levator

Oral view of palate

- Observe midline pulling lateral during /ah/
- Observe indentation in soft palate during /ah/

Nasal view of palate

- Both sides can be seen from one nasal passage
- Normal elevation is complete without air leak during plosives (/pa/, /ka/, /ta/) and fricatives (/sh/, /th/)
- Abnormal elevation is incomplete
- One side lack of complete elevation - gap - especially if viewed from pathway over the middle turbinate
- Bubbles from secretions during phonation - plosives and fricatives
- Can use endoscope as occlusive test - during whistling - knowing diameter of scope
- Also helps patient to realize the extent of injury - often the loss of voice is most obvious injury and may cover up symptoms of nasal leak which can then later become very noticeable after any laryngeal correction.

Lower ganglion branch

Endoscope in upper pharynx, observe midline during a vocal glide from highest pitch to low

- Observe midline raphe which pulls toward strong side
- Observe lateral pharyngeal squeeze from constrictors
- Coincides with pooling/ open piriform sinus

Superior laryngeal nerve

History - frequently occurs after neck surgery

Symptoms:

- Loss of power, loss of upper range
- Loss of singing vocal control or easily matching pitch with singing

Differential diagnosis:

- Bilateral SLN uncommon and symmetric bilateral motion impairment more often represents scar or fixation

Findings:

Vocal capabilities battery:

- Limitation of the extent of upper notes compared to examiner or expected range
- Flattening and poor control in the upper vocal pitch range

Endoscopy

- Vocal cord length changes minimally or not at all during attempted glide up in pitch

Stroboscopy:

- Symmetric oscillations at low pitch while
- Asymmetry of vocal fold vibration at high pitch present because differential tension is most evident when only one cricothyroid muscle contracts.

Recurrent laryngeal nerve

Posterior cricoarytenoid findings

- Arytenoid tipped forward
- Posterior laryngeal fasciculations
- Atrophy of PCA area during sniffing
- No motion lateral of arytenoid during sniffing

Uncommon as an isolated injury, not sure I've ever seen one

Usually the more distal branches of the LCA and TA muscles are also affected

Other notes:

- Vocal capabilities battery: no change
- PCA neurons have a strong tendency to grow back to both the anterior and posterior branches of the RLN
- This usually weights activation of the RLN during respiration toward adduction of the recovered branch

Lateral cricoarytenoid findings

LCA - rotation medially of vocal process and consequently some adduction

- watch respiration for ROM
- watch closure during adduction in slowed down motion for speed of closure and range of closure. Upper portion of arytenoid will cover part of view at end of closure, earlier if the PCA is out on the same side. Low speed very valuable.
- really require topical anesthesia to view posterior commissure during phonation - best view
- Observe for triangle of lateralized vocal process on weak side. Be cognizant of hyperclosure from good cord's vocal process
- Then view at low pitch.
- Compare normal, symmetric posterior commissure with high, medium and low pitch posterior commissures

Other notes:

Arytenoid can be pushed laterally at low pitch in weak LCA, which partly corrects with pitch elevation as CT contractions compensates at higher pitch

View of rigid exam with small "apparently symmetric" posterior gap that changes to an asymmetric gap when viewed up close on flexible exam.

Thyroarytenoid findings

1. atrophy - flexible scope best for visualization
 - a. Atrophy is one of the main factors accounting for diplophonia because different masses tend to vibrate at different frequencies
 - b. Usually requires topical anesthesia to view vocal cord thickness or mass
 - c. Sniffing stretches the vocal cords and augments more subtle atrophy.
 - i. Slow motion helpful for viewing thinning secondary to sniffing
 - ii. Phonation first followed by sniffing also augments.

- d. Can infer atrophy from enlarged ventricle
- e. Inspiratory bernoulli effect if atrophic and if near opposite vocal cord
- 2. fasciculations
- 3. lack of tension

In paralysis (complete paresis), atrophy is often severe and evident, even from above as bowing

- 1. Bowing visualization is augmented by lower pitch
 - a. Must remove cricothyroid compensation in partial paresis - so lower pitch during exam
 - b. If supraglottic squeeze is predominant - implies glottic incompetence more often than intentional hyperfunction
 - i. Topical anesthesia allows passage of endoscope between false cords for a view of cords during phonation
- 2. Augment abnormal vibrations by increasing subglottic pressure. Visualized as
 - a. Flutter - random oscillation, or
 - b. Biphasic - one central node of oscillation with two segments
- 3. Oscillation
 - a. Lateral to axis
 - i. Excursion amplified by low pitch
 - b. Normal cord oscillates nearly symmetrically around central axis
 - c. Normal compensation by cricothyroid muscle leads to obligate falsetto - high comfortable speaking pitch
 - i. Keeps cords near each other
 - ii. Keeps cords in phase or almost in phase (patients tend to avoid diplophonia - especially during an exam)

Other notes:

Vocal capabilities: Husky hoarseness - air leak during obligate falsetto is typical leading to high comfortable speaking pitch, but at loss of sufficient compensation TA weakness is cause of diplophonia because of differential tension.

Reinnervation

- 1. Normal recovery
 - a. Occurs after a minor injury
 - b. Restoration of complete range of motion
 - c. Restoration of appropriate intentional motion
- 2. Synkinetic recovery
 - a. Occurs after a moderate injury
 - b. PCA neurons split reinnervation to anterior adductor muscles and posterior abductor muscles
 - i. On many occasions the vocal cords on endoscopy appear immobile lateral to the midline,
 - ii. or move slightly and movement may be appropriate in direction.
 - iii. Tension is maintained during phonation, rather than flutter.
 - iv. Many times the reinnervated vocal cord is tighter than the "normal" cord during phonation in the vocal underdoer or aged person with bowing of the "normal" cord.
 - v. Perhaps this represents a nearly 50:50 arrangement when abductor and adductor neurons reinnervating LCA and TA.
 - vi. There may also be a mix of abductor and adductor neurons reinnervating the PCA.

- vii. If normal recovery doesn't recur, this is the next best "ideal" spontaneous recovery.
 - viii. That is, it does not require intervention by surgeon.
 - c. Article by Zeitels suggesting that "repositioning" with a temporary injection leads ultimately to more medial positioning in spontaneous synkinetic reinnervation.
3. Dyskinetic recovery
- a. More severe injury
 - b. Overgrowth of PCA (abductor) neurons to LCA muscle
 - i. Vocal process may cross the midline
 - ii. Vocal process may be highly angled toward midline
 - iii. Vocal process may move in the opposite direction of intention
 - i. medial during inspiration
 - ii. lateral during phonation
 - iii. lateral during expiration
 - c. Dyspnea or stridor
 - i. May be light to very strong
 - ii. Not due to weakness, it is hyper reinnervation
 - d. May occur 10 – 20 years after injury
 - i. Often misdiagnosed as unresponsive asthma because of remote interval from injury
 - e. Dyskinetic activity increases with vocal use
 - f. High rate of spontaneous laryngospasm
 - i. Responds to botulinum toxin injection into "paretic" cord.
 - ii. Responds to reinnervation of anterior branch with ansa cervicalis with improved stability and steady tension during phonation
4. Main issue in bilateral RLN injury
- a. Initial symptoms after bilateral injury
 - i. Weak voice
 - ii. Partially out of breath
 - iii. Choking on water
 - b. Symptoms after several months
 - i. PCA (abductor) neurons activate LCA movement toward midline
 - ii. Louder voice, more dyspnea
 - iii. More stridor
 - iv. Increase frequency and intensity of laryngospasm
 - c. Patients often learn to relax TA muscle and/or CTA muscle during inspiration
 - i. Shortens vocal cord
 - ii. Allows central membranous cord to bow laterally
 - d. Botulinum toxin injection
 - i. If placed into LCA muscle, tends to improve airway
 - ii. If placed in TA muscle,
 - i. may precipitate functional airway obstruction
 - ii. bernoulli effect moves membranous cord to midline during inspiration

Tools for an examination

In order to perform a complete laryngeal neurologic examination, the following tools are essential.

- Flexible endoscope with camera
- Digital recording
- Topical anesthesia
- Close exam
- Vocal manipulation

Flexible endoscope with camera

There are two main types of endoscope used in laryngology - rigid and flexible. The rigid endoscope is passed through the mouth and gives a beautiful and detailed, though primarily two-dimensional view of the larynx. The flexible endoscope is passed through the nose. It provides a functional and almost three-dimensional view of the larynx and can bypass many obstructions.

A flexible endoscope, whether the less expensive fiber-optic endoscope attached to a camera or the more expensive integrated video chip endoscope, perhaps even with high definition capabilities, can provide a nearly complete assessment of the three-dimensional anatomy as well as functional motion of the structures innervated by the 10th cranial nerve. In general, the chip endoscope provides a much clearer view and the examiner will learn more using this higher technology. Subtle findings become more obvious.

The two types of endoscope can be used in complementary fashion, but the flexible evaluation is the more essential one.

Digital recording

Recording is essential as so many movements occur over very short time windows. After obtaining a recording, reviewing it slowly, perhaps backwards and forwards, even frame by frame or in very slow-motion, will often reveal details previously missed.

Topical anesthesia

The use of 2% to 4% lidocaine applied topically to the surface of the larynx opens up the possibility of a close view. This can be dripped onto the larynx during phonation leading to a laryngeal gargle. I find that use of 4 mL of 4% lidocaine applied in aliquots of 1 mL each, consistently yields excellent topical anesthesia. Typically the first drip leads to vigorous coughing. After 10 and 15 seconds, the patient is typically able to produce an /i/ sound and maintain it while the topical anesthetic bubbles in the laryngeal introitus. If not by the second drip, then by the third or fourth there is typically nearly complete topical anesthesia.

There are other applicators such as the MAD (mucosal atomization device) which generate and distribute a spray around the pharynx. A thin flexible catheter may be placed to the nose to apply topical anesthesia. In patients with a severe gag reflex, the topical anesthesia may be applied through a 25-gauge needle inserted through the cricothyroid space. The ensuing spontaneous cough will distribute the local anesthesia in both the subglottic and supraglottic areas. Perhaps the most expensive, but easiest way is with a working channel laryngoscope

through which the lidocaine is squirted while the endoscope is held over the vocal cords. Again prolonged phonation while applying the anesthetic generates a laryngeal gargle and more sustained anesthesia.

Notably, topical anesthesia does not alter vocal cord motion.

Close exam

Just applying the anesthesia does not make use of it. The endoscope now needs to be passed into close proximity with the vocal cords. Part of a close exam is the short physical distance of the endoscope to the vocal cords and part of a close exam is observing several cycles of functions such as breathing and also phonation. The endoscope tip can be flexed for a nearly parallel view along the length of the vocal cords.

- Get close
- Observe over time
- Angle the endoscope for review along the length of the vocal cords

Vocal manipulation

In the neurolaryngology exam, the vocal cords should be viewed vibrating at both low and high pitches as well as at both loud and soft volumes. Low pitch and low volume tend to remove compensation for vocal weakness and generate a poor voice. In general, the examiner wants to obtain a video recording during the poorest quality vocal production.

View atrophy

Understand the etiology of diplophonia as an asymmetry in tension, mass or length. A close examination can assess mass, even in patients that appear to have complete closure when viewed from above.

Example of overhead view showing nearly similar size, but flex view shows severe mass difference.

1. Each muscle has a single action.
2. That action can be elicited and to some degree isolated and visualized.
3. Each muscle has an appropriate time to contract.
4. That timing can be compared to the opposite side.
5. Inappropriate timing represents reinnervation.
6. Inappropriate degree of motion represents reinnervation.
7. Patients naturally compensate for any loss.
8. Removing compensation during an exam reveals pathology.

Case histories - excerpted from “Why is there a frog in my throat?”

Paresis – Recurrent laryngeal nerve

Injury and recovery of the main nerve to the larynx

Five years ago, at age 75, Peter McDash was found to have a thyroid cancer. His voice was normal at that time. He underwent radiation therapy and then surgery to treat the cancer and his voice was still normal immediately after surgery. Twenty-four hours later his voice became weak, higher pitched and he lost his vocal durability. Since

then, he cannot compete with any background noise, such as in a restaurant, and that is a significant problem for a talkative person in a talkative family.

He initially sought help from several doctors. One doctor told him that one of his vocal cords was not moving but he should live with it. Another doctor told him that perhaps his thick secretions were straining his vocal cords. He became skeptical that anything could be done to restore his voice, so he has just got along since then without any treatment. Peter's son-in-law previously had a successful voice surgery and finally persuaded Peter to come for a visit.

On first listen, Peter sounds like a frail, 80 year old man with his voice thin and high in pitch, yet he plays tennis every day, suggesting he may not be as frail as he sounds. Thus, there is an incongruity in Peter's story; he sounds frail, but he is not truly frail.

After beginning the endoscopic exam, my first impression is that Peter's right vocal cord is in a position near the midline and not moving, while the left vocal cord opens and closes. Most physicians would agree that he has a paralyzed vocal cord. That may even be exactly what his first doctor said.

Merriam-Webster defines *paralysis* in two different ways:

- complete or partial loss of function or
- loss of the ability to move.

Peter does have a paralysis by this definition. One vocal cord is not moving open and closed when he breathes or speaks, while the other cord retains its opening and closing motions. The injured vocal cord appears (superficially) to be fixed in position.

As a physician scientist, I am ultimately interested in why something is not working because when I understand the why, I can develop a model for a disease and arrive at the most appropriate solution to a problem.

I ask, why did he lose his voice several days after surgery? Why is he hoarse five years later? Why doesn't he and his previous doctors realize that several other symptoms (that he isn't even telling me about and I will learn about later) are also related to the same injury? If we understand the mechanics of the normal vocal cord, why he has lost visible motion and why his voice is poor then we can answer these questions and we can offer solutions. These are questions that his prior physicians didn't completely answer. We can pose more questions and they should still fit the same explanation. For instance, why is his speaking pitch high?

Why did he lose his voice?

Let's start with the first question. Why did Peter lose his voice several days after surgery?

The main nerve that supplies the majority of the muscles of the voice box is called the recurrent laryngeal nerve (RLN). Due to something that happens when we are still not much larger than a ping-pong ball in a uterus, both recurrent laryngeal nerves (left and right) leave the brain, head to the chest and pass around a blood vessel before coming back up to the larynx. On the right side, this U-turn takes place under the collar bone, but on the left side, the U-turn is at the level of the heart.

These long RLNs pass a lot of important structures on their way to the vocal cords: the carotid artery, the windpipe (trachea), the swallowing tube (esophagus), the neck vertebrae, the lungs, the heart and the thyroid gland. If a tumor grows from one of these organs or if a surgeon works on one of these structures, there is the potential for injury to the RLN by stretching or by injuring the nerve's blood supply. If you pull on the cable to your telephone hard enough, you may lose the connection inside the cable. Even when the surgeon just pulls the RLN aside to get where he is going, the RLN can be injured to the point where less signal passes through the nerve.

An interruption of a signal going to a muscle results in a reduced movement of the muscle supplied by that nerve. No signal to a muscle means no movement in that muscle. Superficially that would seem to account for Peter's condition. He has no motion on the side of his nerve injury.

But the answer is more complex. Each RLN, on each side, supplies several muscles including muscles that open as well as muscles that close the vocal cord. When the RLN is injured, the movements required for breathing (abduction) and for making sound (adduction) are often injured simultaneously.

To think further about this let's ask, why didn't Peter get hoarse until several days after surgery? In a long surgery, under general anesthesia, there may be a tube in the windpipe for several hours. It can cause swelling of the vocal folds, so both vocal cords may be swollen enough that with one RLN injured, the vocal fold still working can close against the swollen, immobile vocal cord and make a reasonable sound. As the swelling subsides, the opening between the vocal cords becomes larger and Peter loses his voice because he leaks air through this enlarging gap in the vocal cords.

Why did his voice come back?

Now the double question. Why did Peter's voice come back, yet this recovery was incomplete? Why does he remain hoarse 5 years later?

If the nerve injury is very mild, a reconnection may occur within a few days or a few weeks. The reconnections of the nerves will be appropriate in quantity and the nerve fibers will reconnect with their original muscles. This will get the vocal cord muscles contracting appropriately again. The voice is restored to normal and visible movement is restored to normal.

In anything more than a mild injury, which is the case with Peter, the nerve fibers regrow back through the injured nerve trunk over three to four months and reconnect to the vocal cord muscles. While the nerve fibers have an incredibly strong propensity to find a muscle to connect to, they are indiscriminate or not very choosy. The fibers will hook up to any muscle they encounter that needs a connection. A lot of the connections in the larynx become mixed up because all of the injured muscles are releasing nerve attractants. Some of the fibers programmed in the brain to open the vocal cords inadvertently connect to the muscle that tightens (TA) or to the muscle that closes (LCA) the vocal cord. Some of the fibers meant to close the vocal cord find their way to the muscle that opens (PCA) the vocal cord. With this random reconnection, all of the muscles initially supplied by the RLN may be reinnervated in new proportions or new degrees after the nerve reconnections are complete. Each muscle may receive appropriate as well as conflicting or inappropriate stimulation when the individual wants to make a sound, when he wants to breathe or when he wants to swallow.

For example, the LCA muscle is positioned to move the vocal cord to the midline, such as for making a sound. Let's say that the LCA now receives a signal during breathing in (inspiration) as well as a signal when we try to make a sound. Now it is stimulated all of the time, under all conditions (breathing, phonating, swallowing) and thus essentially remains in a continuous state of contraction. The muscle for opening the vocal cord, the PCA may also receive a constant signal now (during both breathing and trying to make a sound). The vocal cord ends up moving to the middle of the road position with all this stimulation and just stays put. The vocal cord is visibly paralyzed. It doesn't move, but it is not without innervation. The innervation is just continuous rather than intermittently appropriate.

The PCA muscle is the larger muscle, so if enough of these fibers end up going to the LCA muscle instead, the vocal cord ends up in a nearly fixed position somewhere near the midline. Quite often this frozen position is close

enough to the midline that the remaining good vocal fold can get close enough during attempted voice production to set the vocal cords vibrating and create a voice. The good vocal cord can also open wide enough for the individual to breathe in comfortably even though the injured side does not open.

Shortly after an injury, the vocal cord is essentially paralyzed or not moving because it has no signal. Later, it has plenty of signal, but the competing signals counteract or balance each other, preventing effective motion and the vocal cord remains in a nearly fixed position. (This perfect balance only happens when the competing signals are equivalent. Quite often they are not in perfect balance and this will account for the wide variety of symptoms that can be attributed to a neurologic vocal cord injury. I term these disorders “dyskinesia” or dysfunctional motion, rather than “paralysis” or lack of motion.)

Several months after his injury, Peter’s reinnervation fixed his right vocal cord near the midline and his left vocal cord learned to actually cross the midline. Peter’s vocal cords could again nearly close, if not quite as completely as before his injury. He could set them vibrating and his voice returned. Let’s explore some of the details for why his voice remains hoarse despite this closure.

In “Why is there a Frog in my Throat” in the chapter “Two types of hoarseness”, my proposal that hoarseness is only from an air leak or an asymmetric vibration. Lack of vocal cord movement (abduction or adduction) is not a cause of hoarseness. If the vocal cords are fixed next to each other and air is blown through them, they will vibrate and produce sound even though they do not open. We don’t need to describe Peter’s lack of vocal cord movement during breathing to explain his hoarseness, we need to find an air leak or an asymmetric vibration.

I put the scope into Peter, viewing his larynx, noting the obvious lack of opening and closing motion on his right side. But that does not mean there is an air leak. I search for an air leak by having Peter make a sound several times and then I carefully review this process of closing the vocal cords.

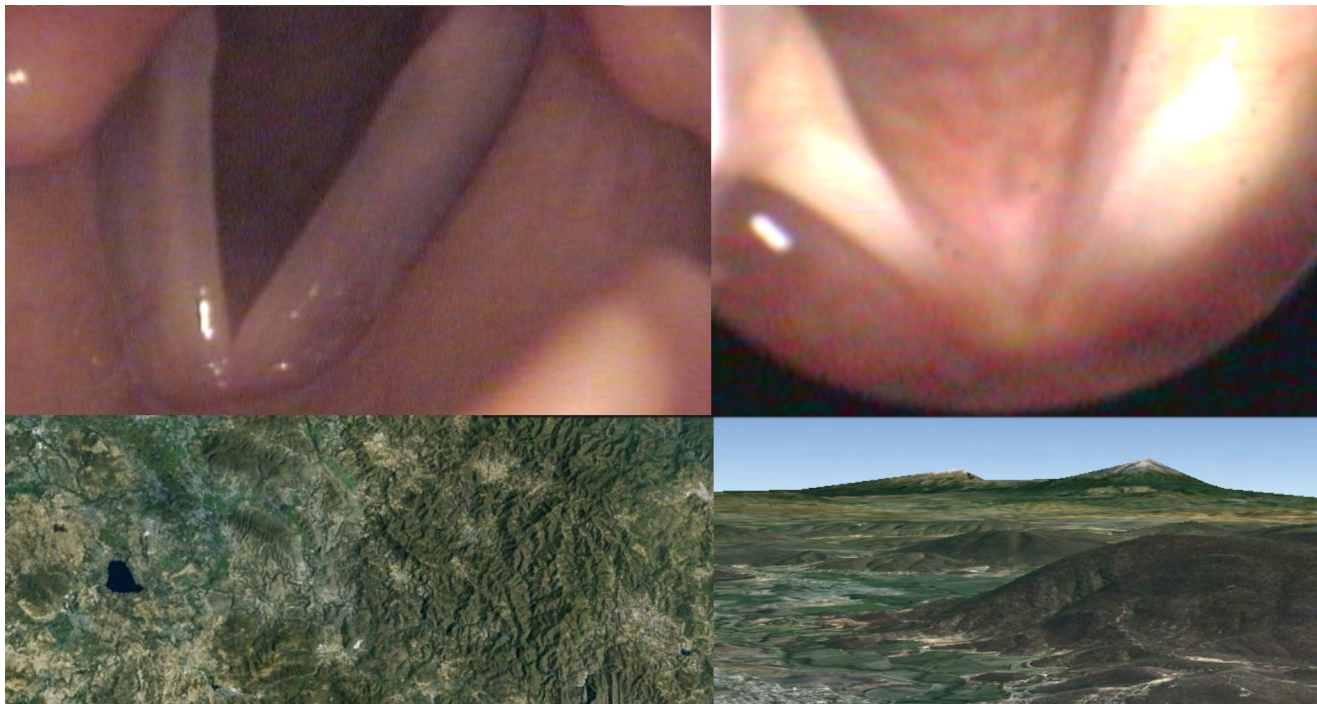
Watching the video in slow motion, initially I see his good side coming completely to the paralyzed side. Then as he starts to build up air pressure beneath the vocal folds, the paralyzed side is weak enough that it actually gets blown slightly sideways, away from the strong left vocal cord during the creation of the sound. A gap opens at the back of the vocal folds at the vocal process. Air leaks along the entire length of the cord.

This air leak affects vocal quality, adding a huskiness to his voice. Additionally, he uses up more air to make each sound, so he gets out of breath quicker with talking than he should. Now we have part of the explanation for the quality of Peter’s voice and for his shortness of breath.

Unknowingly, Peter tries to compensate for this leak. The CT muscle that stretches our vocal cord and puts him into falsetto, is supplied by a different nerve (the superior laryngeal nerve or SLN), so is probably uninjured from the low neck operation. Peter’s CT muscles stretch his vocal cords to try to tension the right side, holding the right cord tight. This supports his weak right vocal cord, but one consequence is that his pitch rises. He is actually speaking in a falsetto voice all the time in order to minimize the gap between his vocal folds. This is the reason for the “frail, elderly man” quality to his voice.

A scope is not a scope

When Peter’s general otolaryngologist physician looked, he announced that one cord was paralyzed, but that the vocal cords were coming together. However, his otolaryngologist likely only viewed the vocal cords from one perspective – directly above and probably far away, blurry, for a very brief period of time, at a single pitch and not recorded for review. At least that is a typical office examination.



Comparison endoscopic views of vocal cords in the same person from above (left), compared to the view with a flexible endoscope inserted all the way to the level of the vocal cords and aimed along their axis (right).

I move the scope closer for a detailed look, until I can align the scope more nearly parallel to the axis of the vocal cords and begin to appreciate the actual thickness of the vocal folds. In cross section, his weaker (paretic) vocal cord is very different in appearance than from above. The paretic cord resembles a linguine noodle in diameter when compared to his healthy cord, which resembles a biceps on a body builder.

Sometimes with a careful slow exam, I can insert the endoscope between the arytenoids to view the entire length of the vocal cords. At other times, I need to drip a little numbing medication on the vocal cords to anesthetize them. If I am not certain about a difference in size between the TA muscles, I have Peter sniff in. Sniffing opens the vocal cords to the fullest extent and pulls them to a long, drawn out position. In this position, his injured cord is less than half as thick as his healthy cord.

When his nerve was injured and even after some portion of it regrew, the RLN could no longer transmit a complete signal all the way to his TA muscle. The TA muscle provides the bulk in the vocal cord. With no signal immediately after the injury, there is no stimulus to the muscle. Over a period of a few weeks, Peter's TA muscle atrophied.

Yet viewed only from directly above, the vocal ligament maintains the illusion that the vocal cord is still wide. The CT muscle also pulls the vocal cord tight as compensation and helps to maintain an apparent straight edge when viewed from above during phonation. These perspective illusions hide a weak, leaky valve.

Sometimes patience rewards the observer. It is difficult for the busy physician to slow down and watch someone breathe. It can take half a minute to watch four breaths and that seems like an eternity to most examiners. I make use of that eternity and watch Peter breathe.

With only one side injured, I can watch and compare the normal side with the injured side. The good cord opens with inspiration (moving air in) and it partially closes with expiration (moving air out). We partially close the vocal folds during expiration to provide resistance to the airflow and keep the lungs more filled with air¹.

On the injured side, the vocal process (the back of the vocal cord) is fixed in position, but the middle of the vocal cord is sucked inward during inspiration (Bernoulli effect) and bows outward during expiration. This injured side is passively reacting to changes in air pressure, while the healthy side is actually tensing lightly during inspiration, so that it is not sucked in by the Bernoulli effect.

Remember, the vocal cords are a valve that control airflow for breathing as well as for making sound. Because his paretic side does not help resist the outflow of air, his lungs do not retain as much air. The alveoli (little sacs in the lung) start collapsing as they empty too completely. Then he absorbs less oxygen from his lungs. The limitation in his tennis game that he attributes to aging is at least partially due to his weak vocal cord not keeping his lungs full. The impact on Peter is that he is more out of breath with physical activity than he should be. He is also more out of breath with speaking, though that is due to the gap between his cords letting more air out during phonation.

In surgery I placed a plastic shim inside Peter's weak and atrophic right vocal cord – a surgery called medialization laryngoplasty or also called a thyroplasty type I. The shim accomplishes two things. It provides bulk to the paretic side so that in cross section, the paralyzed side more nearly resembles the good side in terms of mass. The paretic cord can now maintain tension without the CT muscle pulling on it.

Secondly, while an implant can move both the vocal process and the membranous vocal cord nearer to the midline, that was not necessary in Peter because his partial reinnervation had already put the vocal cord in a good position for closure. However, the implant fixed the cord in that position so that his weak LCA muscle doesn't have to hold up against the stronger left side. The weak but shimmed vocal process can no longer be budged laterally by the good cord or by an increase in air pressure. His good vocal cord doesn't fatigue as quickly with voice use now.

The outcome

Meeting Peter the day after surgery, he told me that when he phoned his daughter after the surgery, she was convinced that she was talking to her brother, not her father! Peter's voice had dropped in pitch. He no longer sounded like a "little old man."

He also noted that he could drink without being so careful. His weak vocal cord was not just impairing his conversation at the cafe, it had been impairing his pleasure of drinking (not alcohol, but any fluid). In retrospect, he realizes that he was choking on liquids more often than he realized. It had just become second nature for him to drink slowly and carefully, so it had not occurred to him to relate this to his nerve injury or to tell me about it. He also found out that he was less winded when he walked. When breathing out, his glottis could partially close now keeping more air in his lung. Lastly, he said that he could speak louder and longer.

He didn't realize that his presenting complaint of hoarseness was actually way down on his list of what he would appreciate by a restoration of his glottic valve. Being less out of breath, not choking so frequently, speaking louder, restoring a more masculine voice, a clearer vocal quality and improved volume all came with supporting his weak vocal cord.

¹ Positive end expiratory pressure (PEEP) – think of a balloon, the more full it is, the easier it is to inflate further. If we let all of the air out of our lungs to the point that the little air sacs collapse, they are much harder to re-inflate than when they are already partially filled. We partially close the vocal folds when breathing out to maintain some back pressure in our lungs which makes filling them easier on the next breath.

To many people (even some surgeons), this is visually counter-intuitive. We actually narrowed Peter's airway at the glottis with this surgery and not only does the voice improve, but breathing through the smaller opening improves symptoms of being out of breath.

Some paresis is visually easy to see such as Peter's nearly immobile vocal cord, but we can be fooled by the patient's compensatory mechanisms that try to tense and close the vocal cords as completely as possible. The underlying reasons for the symptoms can be difficult to account for until we understand the whole problem, and a thorough exam can find them.

Details you may see during endoscopy that represent the "hidden" problems:

- Squeeze of the supraglottis – the false vocal cords squeeze as a compensation for true vocal cord weakness.
- Air leak from a gap between the cords. We can best visualize this gap at low pitches which removes the CT muscle compensation.
- A difference in thickness of the vocal cords represents atrophy of the denervated TA muscle.
- A tense vocal cord will oscillate about its axis. A weak vocal cord will oscillate lateral to its axis.
- On a close exam, fasciculations of the TA muscle may be seen, supporting the diagnosis of denervation.
- During respiration, the Bernoulli effect may in-draw the weak vocal cord.

With 10 muscles in the voice box that can be injured to varying degrees, many nerve injuries are more difficult to sort out than Peter's. Not only can those muscles be injured to varying degrees, but they can reinnervate to varying degrees and there can be cross reinnervation. I am aware that the most commonly used term for this type of injury is paralysis. However, I find that the terms *paresis* and *dyskinesia* best define a neurologic injury rather than the simpler concept of no motion – paralysis. You will hear the term paralysis frequently, but since a lack of motion fails generally to describe why the voice is hoarse, why there is shortness of breath or trouble with swallowing, I prefer to define neurologic injuries of the larynx in terms of which branches of the RLN are paretic (weak) and which are dyskinetic (moving inappropriately). I will more fully explain these two terms in the coming topic "Dyskinesia & paresis".

Peter is now back to playing tennis and enjoying his life because of improved breathing, drinking and speaking.

Paresis – Superior laryngeal nerve

Volume and pitch impairment

Jimmy Nickel works in sales and as a singer-songwriter. He comes in with complaints about singing and loss of projection.

"I noted a sudden pain in my right neck while performing Naphtali in Joseph and his Technicolor Dream Coat. Afterwards it became painful to sing in my upper range so I avoided singing in falsetto. The worst part is that my voice is trashed anytime I have to project. If I use a cell phone, I am just exhausted. If I have to run a meeting, I'm exhausted. I now use a personal microphone with a speaker on my waist just to talk with my wife in the car and avoid projecting my voice. The muscles in my neck are always tight after I have to speak loudly."

Jimmy goes on to relate, "I first travelled to a well-known laryngologist who said I had acid reflux. I took Prilosec for several months without any benefit. Six months later I flew to another city and that laryngologist said the same thing. I took the pill again twice a day, but didn't think it would work and it didn't. Only massage has helped with the neck pain."

Listening to his voice, his upper range is very strained. For a singer, he can only reach about half of a normal falsetto range. He cannot produce soft sounds above G4, fairly low even for a male. For the next six notes, he can only reach them loudly and with great effort and C5 is the absolute limit of his upper range for this trained professional singer.

On endoscopic examination, I ask him to glide up from the lowest notes in his range. As he increases his pitch, there is some rotation of his larynx, with the back of his larynx rotating toward the right side and the epiglottis tipping over toward the right side. His vocal cords do not stretch in length as he tries to go up in pitch. The left cord oscillates about its axis, but the right cord loses tension at his highest notes and oscillates lateral to its axis.

We checked a CT scan of his larynx with specific attention to the cricothyroid joints and there was no visible arthritis nor visible fixation of the joints. By process of elimination, this suggests that the superior laryngeal nerve (SLN) is injured. It is plausible that an accurate electromyogram of the CT muscles would distinguish the injury.

Unfortunately, the SLN doesn't seem to grow back as fully as the RLN. Also, I know of no good treatment to compensate for a SLN paresis. The reduced vocal range seems to be a permanent injury. I cannot say whether the injury to his nerve was a physical one or whether he had some other cause such as a viral infection of the nerve.

Limited vocal cord motion

Vocal cord mobility restriction

There are three types of disorders that might loosely fit under the term paralysis if we use the very broad definition of a lack of motion:

1. After a tube is placed in someone's throat (for example during anesthesia to breathe for them), there may be ulceration between the vocal cords with subsequent scar tissue formation contracting and fixing the vocal cords close together. This may occur to a small degree even with short intubations such as during a surgical case of less than two hours duration. The scarring much more frequently occurs when a tube is left in for many days. The vocal joints may scar enough that the vocal cords cannot move with breathing or speaking. All of the nerves are still sending signals to the muscles. All of the muscles are still trying to respond appropriately. However, the scar tissue holds the vocal cord joints fixed in place.
2. After a surgery in the neck, the recurrent laryngeal nerve may be cut. No signal gets past the injury in the nerve. There is no motion of the vocal muscles with breathing or speaking. The muscles atrophy. There is no motion at all, at least in the acute condition of a complete cutting of the RLN.
3. After an injury to the RLN, the nerve re-grows, but very frequently (because the recurrent laryngeal nerve supplies muscles with opposing action) the individual fibers cross and signals to open the vocal cords go to both opening and closing muscles, so the muscles are activated, but there is no effective motion. The muscles which are stimulated simultaneously compete with each other. There may not even be atrophy of the muscles.

In all three of these scenarios often labeled paralysis, there is a lack of motion, but the etiology is different in each one, so the treatments will need to be different.

Compensation: To add complexity to this, because the function of the 10 muscles in the larynx overlap, if only one muscle is injured, the loss of motion may be partially compensated for by other muscles and on superficial exams, the vocal cords might almost appear to move normally.

Recovery: A nerve may be injured with loss of motion in a muscle, but then regrows. There is partial recovery of motion, but perhaps the muscle cannot do everything it used to do. It may fail to perform its action completely or may only activate sufficiently under stress.

I find the word “paralysis” has too broad of a definition other than to suggest that there is some impairment of vocal cord motion. In general use, this impairment often is neurologic, but not always. If the lack of motion is from scarring, I prefer to call it fixation (and in this book I have described an example of joint fixation in the Autoimmune chapter – see “Arthritis of the laryngeal joints” on page <?> – although trauma may also fix a laryngeal joint).

Beyond that, an injury to a nerve always causes a decrease in function of the muscle it supplies. I try to identify which functions are missing, which muscles have recovered partially and which muscles are competing against each other.

Dyskinesia & paresis

Paralysis – paresis – synkinesis

Mrs. I. Kant Breeve had her thyroid gland removed 30 years ago. She developed a hoarse voice after the surgery (this happens more frequently than some physicians acknowledge²). Her recurrent laryngeal nerve had been injured. Stretched, cut or somehow traumatized, the nerve stopped working after the surgery and one of her vocal cords stopped moving. The cord could not move close enough to the other vocal cord to start vibrating with air flow, so she had no voice for several months.

Gradually her voice improved and eventually it recovered – or more likely it seemed to recover. She had a reasonable speaking voice for 25 years. She could no longer sing nor reach high pitches, but day-to-day, her voice was adequate for work. She accepted that as recovery.

Gradually over the past several years she started having sudden episodes of her breathing being cut off. She would be speaking and suddenly she couldn’t breathe.

Every otolaryngologist who looked told her she had a paralyzed vocal cord, but that she still had “plenty of room to breathe.” Several physicians told her that her episodes of difficulty breathing were from silent acid reflux and they placed her on antacids – the pills didn’t help.

We should ask, do her brief, sudden, “shortness of breath” episodes (dyspnea, usually with stridor) have anything to do with her previous surgical injury? If we wanted to phrase it medically, are laryngospasms related to nerve injuries?

The word paralysis means lack of mobility. It is true that her injured vocal cord does not obviously open or close with each attempted phonation and each attempted breath. There is the implicit assumption that since her vocal cord is not moving normally there is a lack of innervation – an error. The recurrent laryngeal nerve actually

² Why do I say “this happens much more frequently than physicians believe?” Unfortunately, and this occurs much too frequently, she was told to just wait, it will get better. Some people improve in a few days, some in a few weeks and some in a few months. Some people never return to their surgeon having been told “everything will be alright.” They no longer believe their surgeon and seek advice elsewhere. This benign neglect suggests to the surgeon that most hoarseness after surgery is temporary and inconsequential.

All of the hoarseness after a surgery is from some injury during surgery and many of these injuries are nerve injuries. Some injuries are from the endotracheal tube placed in the wind pipe by the anesthesiologist. Since physicians typically don’t look at the vocal folds after surgery and since the patient symptomatically recovers a voice or goes elsewhere, physicians never learn whether the patient should have been put into the “nerve injury” group or the “bruised vocal cord” group and so the surgeon comes away with a false sense of a lower complication rate than exists in reality. The surgeon tells the next patient signing up for surgery, “I rarely injure a nerve.”

has such a strong propensity to regrow that even after cutting out several centimeters of the nerve, it still often grows back to the muscles in the larynx. When the nerve grows back, the major issue is not lack of nerve input, but lack of proper nerve input.

We can say that she is suffering from a laryngeal dyskinesia. Calling her injury a laryngeal dyskinesia implies different findings and different problems than laryngeal paralysis. A dyskinesia may be present whether or not there is any observed motion impairment, though usually there is some impairment of motion. After a nerve injury the problems that result are, to some degree, due to the degree of reinnervation, but even more due to inappropriately directed reinnervation.

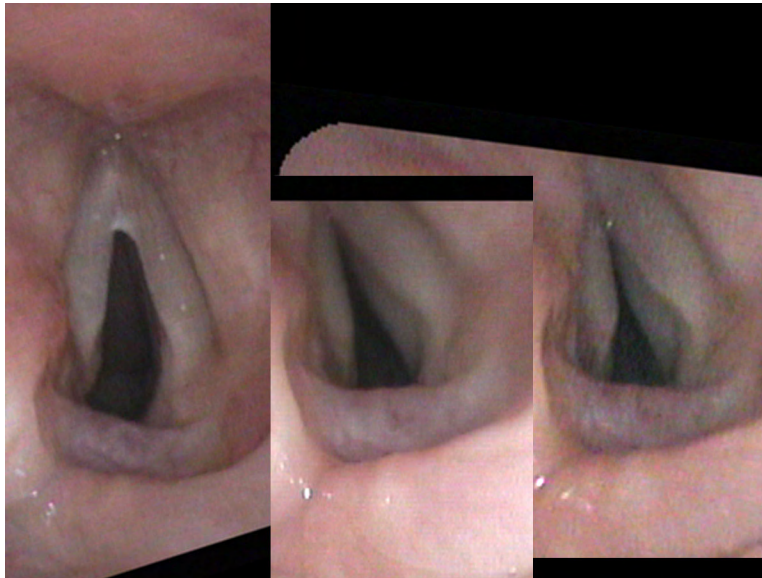
In the ideal world, the injured nerve would regrow back to the muscle it used to control. In the most typical severe nerve injury about half the fibers end up going to their original muscle and the other half go to the opposing muscle. Consequently, the neurologically injured vocal cord appears to be immobile. The brain tells both muscles to contract simultaneously and the net effect is that there is no motion and nothing obvious happens.

That seemed to be the case with Mrs. Breeve for many years, but something definitely changed in recent years. She began having the laryngospasms that cut off her breathing entirely for a seeming eternity (when in reality it was less than a minute, but when you can't get air, time subjectively moves slower).

On her endoscopic exam, the healthy right vocal cord opened and closed appropriately, both during breathing and during sound production. She had quite a strong voice, though I would say that it actually had a too tight, strained quality.

During the ultra-close portion of my endoscopic exam, I touched the left, non-moving or paralyzed cord lightly and it suddenly moved across the midline nearly closing off her airway. It wasn't paralyzed; it could move. It just did not move intentionally and appropriately during breathing or during phonation and it was trigger-happy. With even a small trigger, her left LCA muscle would spasm and move the left vocal process nearly all the way to the opposite cord.

This LCA spasm also increased gradually the longer Mrs. Breeve spoke. With each phonation the right cord would touch the left side and the left vocal process would move further toward the right after each touch. Then, after resting her voice and a number of breaths, the left side would relax back toward its midline, resting position.



Left: vocal cords at initial rest during inspiration.

Middle: after the stimulation of phonating, the left vocal process starts moving across the midline during inspiration.

Right: left vocal process hyperadducting during inspiration and narrowing the airway further after more stimulation.

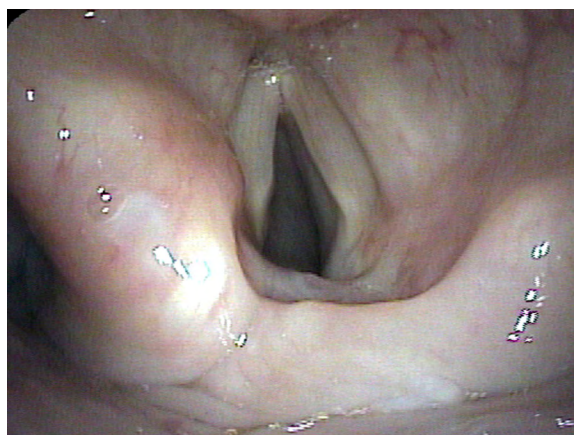
As both treatment and a test, I injected botulinum toxin into her dyskinetic (actually the opposite of paralyzed – hypercontracting), left vocal cord. Specifically, I put it into the TA and LCA muscles – the muscles that tense and move the cord toward the midline and closure.

Two weeks later, the opening in her larynx was larger while she was breathing. She could still make sound, though a little more softly. However, she could breathe better than she had in several years and she had not had any further laryngospasms since the injection. The paralytic effect of botulinum toxin lasted for three to four months and then the nerve regrew again and the left-sided muscles begin to hypercontract again, which she could identify because of the increasing difficulty with breathing. So Mrs. Breeve returned to the office for another treatment every few months. After several injections, she asked if there was something more permanent that could be done.

The RLN splits like a tree into different branches. It is possible to cut only the branches that go to the closing muscles (the TA and LCA) and in effect, that is what we were doing chemically with the botulinum toxin. I suggested a surgery where we would cut the anterior branch of the RLN. Then, to prevent the original RLN from growing back as it had done 30 years ago after her injury, I would route a nerve from one of her neck muscles (the omohyoid) into the cut anterior branch supplying the TA and LCA muscles. If this new nerve sprouted fibers to the muscles before the old branch of the RLN, then she would have nerve input to these muscles during phonation (the omohyoid tenses during phonation). The muscles would bulk up, and even if the muscles didn't have a completely appropriate signal to move open and closed, they would hold tension during phonation and would not tend to inadvertently spasm nor tighten during breathing in.

After cutting the anterior branch of the recurrent nerve during surgery and sewing in the donor nerve, she had a weak voice for a month and then her vocal strength began to return. The left vocal fold ultimately positioned itself near the midline leaving a much larger opening than when I first met her and she could breathe easily. She no longer had laryngospasms that would cut her breathing off and her voice was less tight and strained. She could

close the right vocal cord all the way to the left and had a strong and clear voice without the tight quality she had previously.



Her laryngeal opening during inspiration six months after the denervation-reinnervation surgery to the left side. She has a reasonably open airway with the left vocal cord now stable and set in the midline during both inspiration and phonation and very importantly, no further laryngospasms.

A vocal cord's muscles after a nerve injury are seldom really paralyzed, even if there is no obvious easily recognized movement. The vocal cord doesn't lack movement though it may lack easily visible, intentional movement because of competing contractions. Also, it often has subtle, significant, inappropriate movement. To state it medically, after a nerve injury the vocal fold is more likely to move poorly and inappropriately (dyskinesia) than not move at all (paralysis).

For physicians who approach hoarseness after a surgical RLN injury with benign neglect, feeling that a hoarseness that recovers is inconsequential, they might wish to have their patients examined by a laryngologist to better understand their complication rate. A return to a normal voice might not mean a return to normal function.

Laryngospasm

Acute, sudden spasm

Laryngospasm is the sudden closing of the vocal cords. This is a normal adaptive response to drowning. Let's say you were suddenly submerged under water, breathed in and water began to enter the airway. As soon as the water touched the vocal cords or the lining of the trachea (windpipe) beneath the vocal cords, the vocal cords go into a spasm in a closed position. This stops the movement of air and has the benefit of preventing more water from entering the trachea and lungs. This vocal cord spasm closing off the airway typically lasts for 30 to 60 seconds before gradually opening again (though it seems like forever if you are the one not breathing). Presumably that would be enough time for you to reach the surface. As the vocal cords open ever so slightly, breathing in is very noisy as air passes through the tiny slit (the Bernoulli effect).

Since we don't presumably spend a lot of time drowning, most of us find the inadvertent laryngospasm – perhaps from swallowing while talking – merely a startling inconvenience.

However, there are people like Mrs. Breeve who suffer from frequent, recurrent laryngospasm, some as often as several times per day. My experience is that laryngospasm more than once or twice a year is likely secondary to

dyskinetic reinnervation after a nerve injury. While Mrs. Breeve had an initial severe injury, even a minor nerve injury seems to sensitize the larynx to more frequent and inappropriate laryngospasms.

Whenever I see a patient with frequent laryngospasms, I look for evidence of a nerve injury. If I find it, I consider the use of botulinum toxin even into a vocal cord that is not grossly moving and may have been previously considered paralyzed, since in all likelihood the issue is really dyskinesia or hyperkinetic movement with any stimulation of the injured side.

Sometimes a single botulinum toxin injection will relieve the recurrent laryngospasms. Sometimes the botulinum toxin injections need to be repeated. If repeat botulinum toxin injections are helpful, one can consider a reinnervation procedure with a different nerve supply to block the spasms permanently.

Bilateral vocal cord paresis

A rock and a hard place

Mrs. Abigail Gris awoke from her neck surgery with a weak voice. Dr. Manjoo, her surgeon, says, “Try not to worry, this happens sometimes. The breathing tube in your throat may have caused a little swelling. Just wait, it will get better.”

Sound reasonable? Perhaps plausible, but is it true?

When Mrs. A. Gris didn’t improve after three weeks, she called Dr. Manjoo back. He said, “You better go see an otolaryngologist to have a look at your vocal cords. I have never nicked the nerve to the vocal cords, but it is possible that I stretched the nerve a bit.”

She went to an otolaryngologist who looked in her throat with a scope. He said she had a paralyzed vocal cord, but it looked like it would get better. She should just be patient.

Over the next several weeks, her voice started to recover and in several months it seemed to be back to normal. She and her physicians assumed this was the end of her story.

Do surgeons nick the nerve to the vocal cord? I doubt they make an actual nick or cut the nerve very often. But certain surgeries put the main nerve, the RLN, at risk for injury. I suspect the injury is most often a stretch injury, but the effect of a stretch is initially the same as a cut nerve. Otolaryngologists; orthopedic and neurosurgeons who operate in the neck; endocrine surgeons who operate on the thyroid and parathyroid glands; chest and thoracic surgeons who operate on the lungs; heart surgeons who get near the junction of the aorta with the heart and vascular surgeons who operate on the carotid artery all put the nerve to the vocal cords at risk when they operate near the RLN.

It is also true that an anesthesiologist putting a tube in your throat can also injure the RLN. There is a cuff on the tube placed in your trachea that the anesthesiologist inflates to seal the airway. If the cuff is just barely below the vocal cords and if the cuff is inflated too tightly, the anterior branch of the RLN can be pinched between the cuff and the cartilage of the larynx. The injury will be to the adductor muscles (TA and LCA) and the vocal cords will not close well afterwards.

The anesthesiologist can prevent this problem by ensuring the cuff is far beyond the vocal cords, but more importantly, if the anesthesiologist only puts just enough air in the cuff to stop an air leak, and no more, the nerve should not be injured from excessive pressure.

Mrs. A. Gris did just fine for five years. Inauspiciously, she again developed pain in her neck. Having moved to a new city and state, she found a new neurosurgeon who felt he could help her pain by operating on her neck

again. He chose to approach the neck from the opposite side (I do not know the reason for this decision). Perhaps he heard her good voice and assumed that her vocal cords were working normally and he felt that he could avoid the scar tissue from the previous surgery.

She awoke from this second neck surgery with no voice again. Over a few weeks her voice started to return, but it was still weak when I first saw her. The right vocal cord, previously injured was seen to be in a nearly fixed position near the midline. The left vocal cord, newly injured, could move slightly apart with breathing in. It could move slightly toward the midline with attempted sound production. If she breathed in quickly, the Bernoulli effect would pull in the weak left cord, generate noise (stridor) and make breathing more difficult.

Although her primary concern at this visit was to get a stronger voice back, I asked her to wait and see what spontaneous regrowth would bring. Unfortunately, the regrowth of the injured RLN, while very helpful in a one-sided injury, usually becomes a significant problem when the same regrowth happens on the second side. Over the next several months, her voice did, indeed become stronger as the nerve grew back. Breathing also became more difficult. She saw me again four months after her first visit.

On endoscopy, when she would actively breathe in, the recently injured left cord would actively close. Enough of the fibers that normally opened her vocal cords during inspiration (via the PCA muscle) had grown into the muscle that closes the vocal cords for speaking (LCA muscle) such that, the deeper she breathed in, the more strongly the left cord closed.

She now has a dyskinetic reinnervation. The vocal cords are not moving a lot, so superficially they might appear paralyzed, but on our close examination, this small, but very significant movement closing her left vocal cord is actively shutting down her breathing. Six months after her second RLN injury she has to avoid or minimize any activity or she cannot breathe well. The more she tries to do, the less air she gets. Her voice has become loud enough now, although it has a tight, lightly spasming quality. She lacks fine control over her voice.

Her options have become very limited. The simplest surgical solution is to place a tracheostomy tube. A tracheostomy is a hole in the neck with a plastic or metal tube inserted to keep the hole open. This allows breathing in and out through the neck bypassing the vocal cords. A one-way valve can be placed on the tracheostomy and air can enter the trachea on inspiration through the tube, but is then directed out through the vocal cords for generating sound or voice on expiration. Then she can both breathe and speak.

There are other options, such as permanently cutting or partially removing one of the vocal cords or tying one of the vocal cords out into a permanent open position. One can partially or completely remove the joint (arytenoidectomy) that opens and closes one of the vocal cords to enlarge the airway.

However, there are several problems with creating a fixed opening into the windpipe. The voice becomes softer the larger the opening. Although she will get air in, more of the air will leak out as well, and she will likely be more out of breath with speaking and activity. It is not a restricted breathing, but rather it is an "out of breath" sensation. Lastly, since the vocal cords are a valve, not only for breathing and voicing, but for swallowing, the larger the opening into the windpipe, the more that liquids and food go down the wrong way. So coughing while eating increases and pneumonia becomes more common, since food in your lungs is not very healthy.

Healthy vocal cords are an active valve that dynamically open and close to regulate airflow and food flow. With a fixed valve or a malfunctioning valve, such as with her bilateral RLN injury, one has to make choices between a completely closed valve (no air movement, so not very functional), a completely open valve (no sound production and lots of food in the lungs, so not very functional), or an in-between, leaky valve. There are no good choices, just bad ones with a lot of compromises.

Mrs. A. Gris started by getting a tracheostomy. She really didn't like this option, but it very quickly allowed her to breathe very easily. In order to see if we could get the tracheostomy tube out, we tried to cut the anterior branch of her recently injured nerve and grafted a new nerve onto that branch. It opened her airway slightly, while still preserving her voice, and it allowed her to function with a one-way valve on the tracheostomy tube more easily.

She still wanted to try and get the plastic tube out of her neck, so we started to remove portions of her left vocal cord and left arytenoid cartilage. She could now plug her tracheostomy tube for extended periods of time and her voice was a little softer, but there were times with exertion where she had to open the tracheostomy tube to get enough air. Consequently she could not do without the tracheostomy tube completely.

She started to get recurrent pneumonias from small pieces of food and liquid that would get through her now partially open vocal cords. We had reached a limit to what I could do. If I opened her vocal cords further, she could have the tracheostomy tube removed, but her voice would become more of a whisper and the pneumonias would likely increase. She appears to be stuck with a tracheostomy tube in order to breathe.

If you have ever have a RLN injury, before considering any additional surgery on your neck or chest, you should have a good laryngeal exam to assess the risk of an additional surgery. Even with a normal voice, it is possible to have one vocal cord that does not move well or at all. If that happens to be the case, then before another surgery near either RLN, an updated exam will allow you to understand the incredibly high risk to operating anywhere near the remaining uninjured nerve. Operating near the already injured nerve is a small risk. Operating near and injuring the nerve on the opposite side ends up as a significant, life-altering event.

I consider this analogous to our vision. We have two eyes which offers some redundancy. Losing one eye means a loss of depth perception, but we can still see to read, drive and look at the smiles on our friends' faces. But losing the second eye leads to complete blindness. We are surely still alive, but we completely alter the way we live. The RLN supplying the vocal cords is the same type of partially redundant system, very forgiving when one side is injured, unforgiving if the second side is injured. Mrs. A. Gris could tell you how life altering this injury is.

Laryngeal dystonia

Adductor spasmodic dystonia

At age 83, Effie Forte reported one year of effortful speaking. If she shouted for awhile, she could continue to speak with effort. After resting her voice though, it was very difficult to get her voice started again. She spoke in short, tight bursts when describing her problem.

On endoscopy, I noted that she squeezed her larynx tightly from each side every time she tried to produce a sound and that sound would be cut short by the squeeze. Except for the severe squeeze, her vocal cords appeared normal and using stroboscopic light, they vibrated normally for the brief period of each vocal effort.

I felt that her diagnosis was adductor spasmodic dystonia (also commonly called adductor spasmodic *dysphonia*). Adductor spasms of the vocal cords mean that the vocal cords over contract, irregularly closing tightly when trying to phonate. It is a focal dystonia, which are spasms of a single muscle or a group of muscles. Spasms tend to be irregular and occur during intentional use. They usually do not occur at all when the muscle is at rest.

While a tremor is also an over-contraction, often occurring only during intentional movement, a tremor has a regularity about it. As one muscle over-contracts, the opposing muscle takes over and an oscillation starts as each opposing muscle contracts in turn. These contractions tend to be regular and occur typically about 4 times per

second. They are perhaps most well visualized when a person with a hand tremor tries to pick up a coffee cup. If the cup is full, the coffee slushes out each side of the cup. We call this an intention tremor since when the person is not trying to do something, the tremor may become unapparent. It occurs with intentional movement.

Spasms of dystonia also occur during an intentional movement, but there is no counteracting muscle contraction. Spasms can be a nearly continuous contraction or a very intermittent and irregular contraction.

In the larynx, spasm of the closing muscles during phonation, *ADductor* spasmodic dysphonia (AD SD) is the most common type of laryngeal spasm and causes the voice to cut off during speech as the vocal cords clamp shut. *ABductor* spasmodic dysphonia (AB SD) is less common and when trying to phonate, the muscles inadvertently open, causing breathy breaks in the speaking voice.

AD SD is one of the few laryngeal disorders that is often more easily visualized during speech than during a steady sound. A person with AD SD can overpower the closure of the cords and emit sound, but this is very fatiguing. Mrs. Forte had such a tight squeeze that it was like a single strong spasm every time she spoke.

I recommended botulinum toxin injections to weaken the muscles closing her vocal cords. No one knows what causes a dystonia, though I suspect the problem is in the brain, not in the nerve endings. However, for now, the best available treatment seems to be partially blocking the nerve input to the spasming muscles. It seems reasonably safe to do so with botulinum toxin which wears off if the result is not beneficial. The wearing off is also a downside as the beneficial effect wears off and the injection needs to be repeated in three to six months.

Mrs. Forte was not very excited about needing repeated treatments several times per year. The injection is fairly quick, but unlike a pill for a disease that can be taken at home, most people find it difficult to place a needle into their own neck, find the vocal cord muscle and insert the medication. Thus, the treatment requires a trip to the doctor, a higher cost because you have to pay the doctor as well as pay for the medication and, while brief, there is the not terribly pleasant idea of having a needle inserted into your neck.

She wanted a cure, not a treatment, but agreed to try the medication. She was able to speak more freely after the treatments for three to nine months depending on the dose. However, after three treatments she still sought a cure, not a treatment. I felt we learned something by the treatments. She was improved after each treatment, suggesting at a minimum that the diagnosis of AD SD was correct.

I didn't hear from her for awhile, then about 18 months later she returned. Speaking was very effortful again. Her voice was worse than before. On endoscopy, now her vocal cords had lost some of their white color. Gradually I learned that in the interim she had gone to a general otolaryngologist who told her that she had Reinke's edema. He had not taken any photos, but he had taken her to the operating room and had "stripped" her vocal cords to remove the edema. Her voice was worse after the surgery and had not improved with time as the surgeon had predicted.

"Reinke's edema" is a condition also known as "smoker's polyps", which tobacco smokers who talk a lot develop. In fact, only tobacco smokers who talk a lot develop them. There is something about the combination of smoke and the vibration of the vocal cords that causes the edema. Besides that, the only complaint patients with smoker's polyps typically have is "Do I have a cancer?" The change in their voice is a deeper pitch because the vocal cords are thicker. Thick cords vibrate at a lower pitch just like the thicker strings on a guitar or piano. However, the females and the males I have met with smoker's polyps not only aren't concerned by their pitch, they often like their deep pitch, even the women who are so deep they are mistaken for men on the phone. They just say that everyone who knows them recognizes their voice. If they don't have cancer, they don't want to get rid of their smoker's polyps. Imagine a female jazz club singer and you likely have the sound of smoker's polyps in your mind.

Mrs. Forte did not talk a lot, did not ever smoke tobacco, did not have a deep voice and I had photos of her vocal cords from one-and-a-half years ago that were normal in terms of their structure.

One of the problems for physicians is that their word carries as much weight as a photo or video recording. We quite often answer only to ourselves. Since the physician who diagnoses the patient also treats the patient, the diagnosis and the treatment always seem to line up in a report. I really don't know how to explain this particular physician's diagnosis of Reinke's edema in a women who did not ever smoke and who on my video recordings had normal vocal cords only 18 months ago – error, jealousy, wanting to please the patient? Now the patient has two problems, stiff vocal cords (from the stripping) and spasmodic dysphonia. She is now in a tough position.

Botulinum toxin injections may not be the only treatment for some laryngeal dystonias. When the spasms involve the adductor branches of the recurrent laryngeal nerve, then surgically cutting the adductor branch of the RLN and hooking up a sacrificial nerve from another neck muscle (ansa cervicalis) is also an option. This procedure is called a DEnervation-REinnervation surgery (DeRe) or a selective laryngeal adductor denervation (SLAD) (physicians love complicated, if accurate, names and their associated abbreviations). However (there is often a however), her insurance will not cover this procedure, labeling it “experimental.” It is also expensive.

She has public health insurance designed for the poor. Interestingly, the insurance paid for the vocal cord stripping (remember the diagnosis and the treatment lined up on a list at the insurer so approval was given). It would cover the botulinum toxin injections with enough paperwork on my part, but it wouldn't cover the DeRe surgery.

She continues to live, speaking with great effort and now with a poor quality voice when she is able to get sounds and words out. Many times I ask myself, what is the value to a patient and to society of a correct diagnosis?

Abductor spasmodic dystonia

In his mid-fifties, Bob Icon has had to reduce his preaching schedule to a minimum. For the past 15 years his voice has become progressively more undependable. His voice fades out, breaks up and sounds unsteady. It requires a great deal of effort to speak. The first ENT he saw diagnosed vocal polyps and sent him to speech therapy. However, the therapist told him that his voice sounded like he had abductor spasms. He went to a second ENT who tried injecting botulinum toxin using a ultrasound for guidance, but each time it made his voice breathy for awhile without improving his voice. At one point, he had a cold and his voice improved temporarily during the infection. He was sent to a neurologist who gave him clonazepam. This made him sleepy and made his snoring worse and didn't seem to help his voice.

Abductor laryngeal dystonias are spasms of the opening muscles of the larynx, the PCA muscle. The patient has “breathy breaks” in the voice while speaking. This type of dystonia may be treated with botulinum toxin injections into the PCA muscles.

Although he states that one physician tried to inject the PCA muscles, it sounds like she probably never actually placed the medication in the correct muscle based on Bob's description of the injection. He described what sounded like a TA muscle injection and he had breathiness after the injection – a typical side effect of a TA injection. Since the PCA muscles open the vocal cords for breathing, they will not open the vocal cords as far if they are weakened by botulinum toxin. Vocal cords that cannot open well can impair breathing, and with the vocal cords closer together the voice should be stronger. If too much botulinum toxin is placed in the PCA muscles, the patient will make a noise with every breath inward because they cannot open the vocal cords far enough apart to

prevent vibrations from the Bernoulli effect. If too little botulinum toxin is placed there will be no effect. He had neither of these effects.

The fact that he had a weak, breathy voice after the injections suggests the botulinum toxin was actually in one of the vocal cord muscles that tighten the vocal cords, such as the TA or the LCA muscle. Whether that was intentional or inadvertent on the part of the physician, I couldn't know with the information I had.

I discussed with the patient that different physicians use different techniques for injecting the PCA muscles. Some physicians try to pull the larynx forward, which is uncomfortable, and pass a long needle behind the larynx. I utilize an EMG and I pass the needle straight through the front and out the back of the larynx so that I know when I am encountering the PCA muscle by the feel of the cartilage and the EMG signal. I also told him not to expect breathiness as a side effect, rather there should be some mild restriction in his breathing, possibly noisy breathing.

Part of my diagnostic process is to place the botulinum toxin and see the patient back in two weeks and see if the intended muscle is weakened. If the muscle is not weakened, then we would try a higher dose. If the patient is not improved, despite weakening the intended muscle, then we would need to rethink the diagnosis. He was, however, improved and has resumed his preaching as well as continued with the injections.

Respiratory dystonia – inspiratory

For several years Mr. S. Tate Seller has noted pain in his shoulders and neck, and for two years he has noted difficulty talking and breathing. When he tries laying down to sleep, he struggles for about two hours before falling asleep because of difficulty breathing in. His breathing seems better in the morning, but as soon as he starts talking or moving, the difficult, noisy breathing returns. His voice is normal, but his breathing is very noticeable, gasping between words, especially on the phone. He is an estate broker, and when people hear him gasp he loses their confidence as well as the sale. He cannot even sell enough to pay for health insurance anymore. When stressed, his breathing is more restricted and he has to continuously clear his throat.

Chiropractic treatments and massage help reduce the neck and shoulder pain temporarily, but have not improved the noisy breathing. His family doctor diagnosed him with anxiety (try not being anxious when every breath is noisy and difficult). He lost weight, but the anti-anxiety pills did not improve his breathing. He was told it was due to allergies. Allergy medication did not help, but clonazepam, a muscle relaxant, did improve him a bit.

On endoscopy, I note that he can speak easily and that his larynx is smooth and stable during phonation. However, when he attempts to breathe in, instead of opening, both vocal cords appear very unsteady as he tries to breathe in, spasm closed and create quite a bit of noise during inspiration. The vocal cords never really open anywhere near normal except briefly during a cough.

While the more common dystonia of the adductor muscles occurs during attempts to speak, his spasms occur during attempts to breathe. Dystonias typically occur during a task or intended muscular activity. They are an irregular activation where the muscles contract much more than they are supposed to. Other perhaps more familiar examples include torticollis, where a large neck muscle contracts pulling the head sideways; blepharospasm, which causes the eyelids to nearly continuously blink and writer's cramp, which occurs in the hand as fingers cramp up while trying to write or play an instrument. Mr. Sellers's LCA muscle is spasming when he tries to breathe in, an exhausting condition. By the time I see him, he has to sit up in bed struggling for every breath until pure fatigue renders him asleep.

I inject botulinum toxin into the closing muscles of his vocal cords and one month later when I see him, he has stopped all of his nasal inhalers and all of his allergy medications because they were unneeded. Speaking is easier, breathing is easier, though he still audibly gasps between words. Sleep is also coming on easier.

On future injections, we place larger doses of botulinum toxin and he breathes better, falling asleep easier. After a number of treatments, between injections he never returned to as severe a condition as when I first saw him, even if he goes for a long period between injections. There seems to be some partial permanent improvement after several injections. Mr. Seller receives injections about every six months.

Respiratory dystonia – expiratory

Mrs. Tock complains that she has too much phlegm, a rather common complaint in an ENT office. During endoscopy, there is no excess of secretions but there is an unusual movement of her vocal cords during breathing. Her vocal cords open to take in air, but then close prematurely and sit against each other shortly after she breathes in. Normally, vocally cords close slightly during expiration (breathing out) to provide some resistance and keep the lungs from collapsing. Hers are closing most of the way together, such that the flexible part of the vocal cords are completely together. I find that many times the sensation of excessive mucous production relates to only a small amount of mucous which ends up sticking to the vocal cords. For Mrs. Tock, with the membranous vocal cords sitting against each other she senses the touching as phlegm build up, but it is actually each vocal cord touching the other that she is sensing. There is no extra mucous. She has spasms of her LCA muscles occurring during expiration, so this is a respiratory dystonias of expiration.

An injection of botulinum toxin into the adductor muscles softens her voice, but also allows the vocal cords to rest apart during breathing and relieves her sensation of phlegm build-up for several months.

Other dystonias

Muscles surrounding the larynx, muscles supporting the larynx and muscles involving the speech tract above the speech line (pharynx, tongue, lips, jaw) may spasm and impair airflow and thus impair voice and speech. These dystonias may interrupt or alter speech and so may sound similar to some of the above laryngeal dystonias that involve specific vocal cord muscles. Some regional dystonias respond well to botulinum toxin injections, particularly the supraglottis. With muscles more closely involved in swallowing, side effects from botulinum toxin weakening swallowing can make treatment of the muscle spasms not worthwhile for the patient.

Summary of dystonias

Dystonias can involve a single muscle or a local group of muscles. They occur as a spasm during attempted voluntary movement, effectively an inappropriately strong movement when not desired. Whether the spasm occurs during breathing or speaking, they also may involve some of the nearby muscles in the neck which are recruited to help move air through the tightening vocal cords. This caused Mr. Seller to have neck pain from chronic muscle tightness. This neck pain also improved after each laryngeal treatment.

Dystonias of the larynx most frequently occur with phonation, typically the TA or LCA muscles, interrupting and cutting off the voice and consequently interrupting speech. Spasms of the PCA muscle during phonation cause inappropriate opening, which in turn causes breathy breaks in sound production. Depending on the timing of spasms of the TA or the LCA muscles or both, interruptions can also occur with breathing in, cutting off the airway as with Mr. Seller's or with breathing out as with Mrs. Tock.

Vocal tremor

Highly regular inappropriate movement

At 87, Elizabeth Longlife comes in with her children. The children complain that Elizabeth is getting more difficult to understand over the past five years. She sounds unsteady to them. Ms. Longlife states that she is not bothered by her voice but came in at the request of her children.

Unlike the irregular spasm of AD SD, a vocal tremor is a regular oscillation of the larynx giving the voice a very shaky or unsteady sound. Frequently, a vocal tremor does not bother the person with it. Mostly it bothers those who have to listen to and understand the person with the vocal tremor.

Generally, a patient with spasms senses a great deal of effort and feels they are fighting against the spasms. A patient with tremor does not sense any significant increase in effort with making sound. Her voice is just unsteady.

Vocal tremor can be mitigated to some degree with medications that work for other types of tremor. The most common medication is propranolol, which will tend to decrease the intensity of the oscillations but not eliminate them. Mrs. Longlife was happy to know that she didn't need any more medications unless the tremor was to the point that it bothered her.

Neurologic combinations

Spasms and tremor are associated

It is not uncommon to have more than one of the above types of tremor or spasms. About 10 percent of people with AD SD also have a vocal tremor. A person may have both adductor and abductor spasmodic dysphonia. A patient may have a vocal cord dystonia and a neck dystonia. In a person with a mixed neurologic voice disorder, I typically begin treatment for one of the disorders at a time so that both the patient and I may decide how effective each treatment is alone before combining them.