Dr. Linda Yin:

Hi there. Welcome to another episode of ENT in a Nutshell. My name is Linda Yin, and I am joined today by laryngologist, Dr. Alex Gel bard. Dr. Gel bard, thanks so much for being here.

Dr. Alex Gelbard:

Thanks for inviting me, Linda. I'm excited to talk.

Dr. Linda Yin:

Great. Today, we're going to talk about adult laryngotracheal stenosis, otherwise known as LTS. Before we jump into the presentation of this disease, I think we should define some terms for the listener, because the nomenclature can be really challenging and confusing. So, what is LTS?

Dr. Alex Gelbard:

Well, when I use the word LTS, it's an umbrella term describing several disease process with a common physiologic endpoint of lamiltory restriction. And they all come from luminal narrowing at the level of the larynx, or the subglottis, or trachea. And they manifest as fixed extra thoracic restriction of pulmonary ventilation. And given the overlap of LTS with a lot of intrinsic pulmonary disease, as well as the contribution of systemic conditions, most successful high volume programs in peds and adults make multidisciplinary care of airway disease a cornerstone of treatment.

Dr. Linda Yin:

You mentioned that LTS is really an umbrella term. So, what specific disease processes does it actually contain or describe?

Dr. Alex Gelbard:

Well, although most airway stenosis appears similar on anatomic imaging and clinical exam. Different mechanisms of injury are associated with different rates of longterm, tracheostomy dependence. So, trying to differentiate the etiology, I think is important for helping people understand the outcome and risks down the road. So really, we divide LTS into some broad categories: one of them being iatrogenic, or disease that develops following intubation or tracheostomy; another would be auto-immune, so things that happen in the setting of a collagen vascular disease, the biggest one being granulomatosis with polyangiitis, formerly known as Wagner's; and then another small disease that we'll see would be relapsing poly-chondritis.

Dr. Alex Gelbard:

And then, the other big category would be idiopathic subglottic stenosis, where disease arises without a known cause. Some other more uncommon causes of laryngotracheal stenosis include trauma and radiation, or infection. Overall most large series from real high volume centers around the world suggest that about 70% of disease results from fibrotic contracture after endotracheal intubation, about 15% from ISGS, and about 15% from granulomatosis with polyangiitis. So, the remainder radiation, trauma, infection, burn occur with much lower frequency.

Dr. Linda Yin:

Okay. Let's move on to the presentation section now. So, how does a patient usually present to you with LTS?

Dr. Alex Gelbard:

Well, most common presenting complaint is dyspnea, or shortness of breath, but people can also present cough or dysphonia, and rarely they can have trouble swallowing.

Dr. Linda Yin:

When you're evaluating a patient in clinic who is presenting with symptoms of dyspnea or even respiratory distress, what sorts of goals do you have and what are you focusing on?

Dr. Alex Gelbard:

Well, the first thing you're trying to do when you're evaluating somebody with a complaint of dyspnea is triaged patients that need urgent intervention to prevent real eminent death. And those are the people that you'll see in the emergency room, and really need rapid action. And that's probably a whole nother discussion. You're also trying to determine patients who have symptoms, but without severe ventilatory impairment and they can be safely evaluated in clinic.

Dr. Alex Gelbard:

And so, first thing you're trying to do when you see somebody is make that decision, "Is this an emergency or do I have a little more time to try to sort this out?" And then those people that make it to clinic much like a cancer patient in the initial evaluation, I want establish the etiology of their dyspnea and in the process, localize disease to a specific anatomic sub-site, "Is this a problem in the subglottis, in the glottis, in the super glottis, or in the trachea?"

Dr. Alex Gelbard:

I'm trying to define the type of obstruction. Is it primarily mucosal scar, or is there a collapse of the collagenous airway framework, or both there? Because the answer to this question has pretty significant implications for treatment. And then once they're diagnosed, I want to stage their physiologic and functional impairment to help calibrate the intensity therapy and articulate the degree of risk associated with the different therapeutic options out there for them.

Dr. Linda Yin:

When you're seeing a patient in clinic who has signs and symptoms that might be concerning for LTS, stenosis is on the differential. But what else is on your differential diagnosis?

Dr. Alex Gelbard:

Well, there can be benign epithelial derived tumors, so papilloma or cysts; you can have benign non-epithelial tumors, schwannomas neurofibromas, et cetera; you could have malignancy derive from that, epithelium, ie. squamous cell carcinoma; you can have mesenchymal malignancies like a chondrosarcoma; or you can have salivary gland malignancies, like an adenoid cystic carcinoma. You can have intrinsic pulmonary disease, so asthma, interstitial lung disease, idiopathic pulmonary fibrosis. Or you can have an infection bacterial, fungal, or microbacterial. Those are the things that I'm really trying to make sure I don't miss.

Dr. Linda Yin:

Now, we would typically talk about pathophysiology here, but the pathophysiology of this disease can be a little bit confusing. So, I think I'm going to jump ahead a little bit and let's talk a little bit about

clinical workup first. So, how do you establish the diagnosis of LTS and are there any diagnostic criteria that's used?

Dr. Alex Gelbard:

So, the core elements of diagnosis include history, physical examination, laboratory studies, and then other adjuvants that are helpful, pulmonary function testing, and radiologic studies.

Dr. Linda Yin:

Okay. Now, how about the physical exam? What components are you focusing on there?

Dr. Alex Gelbard:

Well, I think the physical exam is really important, and it's critical to carry out the exam in a reproducible, orderly way. And understanding the pathophysiology of extra and inter thoracic obstruction helps avoid some diagnostic mistakes. One thing that's helpful is really listening to the patient's respiratory sounds, as this will provide some real good information to the severity of their distress, and help you localize their obstruction.

Dr. Alex Gelbard:

Identifying the site and cause of obstruction is really facilitated by the quality of the pathologic sound. So, is there stridor, is there stertor, or is there wheezing, and the phase of the respiratory cycle during which it's most pronounced, is it inspiratory or expository? Another thing is, nasal inspiration really forces maximal vocal cord abduction, really opening the larynx. So, I'll always ask patients to take a big breath in through their nose during my assessment to really delineate how much is the larynx contributing to any sound that I hear, and try to give the best possible examination of their airway with their larynx maximally open.

Dr. Alex Gelbard:

So, any airway obstruction generates respiratory sounds from the rapid turbulent air flow through the narrowed segment of the respiratory tract. And depending on the severity, and along with other symptoms, you can see nasal flaring, supraclavicular attractions, sternal retractions, intercostal and subcostal retractions. And these help indicate the intensity of the respiratory distress.

Dr. Alex Gelbard:

No other parts of the physical exam, after just a general observation, include the oral cavity. So, I want to know about the mouth opening as a potential laryngeal exposure for laryngoscopy. Is this going to be a difficult exposure? Does some evidence of obstructive sleep apnea or a phenotype consistent with obstructive sleep apnea contribute to their symptoms? I'm also looking at the nose, and I want to look for a mucosal disease in the nose. Something like crossing the septum or septal perforations that would heighten suspicion for vasculitis. And additionally, nasal obstruction, if you find it will exacerbate laryngotracheal obstruction, and it's worth investigating.

Dr. Alex Gelbard:

So then, after these things, I move on to trans nasal fiberoptic laryngoscopy, and this is a real integral part of the exam. First question I'm trying to address is, "Does the glottis move?" And then I want to know, "Is the subglottis patent, or does the mucosa appear inflamed?" And autoimmune conditions can

really affect the glottis and subglottis, so that a strong history of intubation or trauma, laryngeal impairment, or very high subglottic disease, what you might term "infraglottic involvement" really raises suspicion for me of an auto immune pathology.

Dr. Linda Yin:

Speaking about autoimmune pathologies, are there any laboratory studies that you routinely get as part of your workup for LTS?

Dr. Alex Gelbard:

So, these include labs to rule out collagen vascular disease manifesting as airway obstruction, ie. granulomatosis with polyangiitis, and this is most commonly c-ANCA testing, or cytoplasmic antineutrophil cytoplasmic antibodies. And these are commonly target against proteinase three, or myeloperoxidase. Now, if this is positive, it's strong evidence that vasculitis is playing a role in their disease. If they're negative, it helps push you away from a autoimmune diagnosis. Although, there still are some people who will have a vasculitis and be ANCA negative. But in large part, this laboratory study can be pretty informative.

Dr. Linda Yin:

How about pulmonary function testing? What is the role of pulmonary function testing in the diagnosis of this disease?

Dr. Alex Gelbard:

Well, I don't get pulmonary function testing for every patient. But when the diagnosis is complex or there's a question about the contribution of intrinsic pulmonary disease, I think PFTs can be really helpful. So, LTS has a fixed, extra thoracic obstruction pattern on PFTs, with the flow volume loop diminished in both the inspiratory and expiratory phases of disease.

Dr. Alex Gelbard:

And this is notably different than inter-thoracic variable obstruction, which typically affects expiration; and extra thoracic variable obstruction, which typically affects inspiration alone. There's been a lot of objective indices that have been evaluated to aid in diagnosis, and these include the expiratory disproportion index, which is the ratio of FEV1 to peak expiratory flow rate. And values greater than 50 have a sensitivity of about 96% and a specificity of 94% in differentiating between fixed extra thoracic stenosis and non stenosis.

Dr. Alex Gelbard:

Another study that's utilized is the maximum voluntary ventilation, and this is the amount of breaths you can take in a prescribed period of time. But one of the most important values to me is the measurement of the alveolar capillary surface area available for gas exchange, which is measured via the pulmonary diffusion capacity for carbon monoxide. What we call the DLCO, and reduction to this value indicate intrinsic pulmonary disease. And for these patients, if they have a very low DLCO, even if I want to correct their extra thoracic obstruction, I'm not going to relieve their subjective dyspnea. So, knowing this ahead of time, I think, is very important.

Dr. Linda Yin:

And we talked earlier about imaging. So, what imaging modalities are helpful in the workup for LTS?

Dr. Alex Gelbard:

I often find that thin cut, non-contrast CT is helpful to localize the side of anatomic obstruction, as well as define the extent of disease. It doesn't work superbly well to delineate laryngeal pathology, but it's very effective for subglottic and tracheal disease. And if there's a question about variable obstruction, ie. dynamic instability, tracheomalacia, we'll obtain a specific protocol with inspiratory and expiratory phase images obtained.

Dr. Linda Yin:

Okay, so now we've done this whole workup and diagnosed LTS, how do you know how severe it is? And is there a staging system that we can use?

Dr. Alex Gelbard:

The real question is, why are formalized staging systems important? And adult LTS comprises a wide array of conditions that require precise preoperative assessment to help you individualize surgical treatment for everyone. And historically, the data about operative management failed to report complete information on really relevant issues. And these would include vocal fold mobility before, cicatricial glottic and super glottic involvement, additional tracheal damage, stenosis malacia related to an airway prosthesis, secondary airway lesions, obstructive sleep apnea symptoms, swallowing trouble, severe systemic illness like gastroesophageal reflux disease or eosinophilic esophagitis, as well as medical comorbidities.

Dr. Alex Gelbard:

So, the literature had a lot of case mixtures with various conditions and it made it difficult to compare postoperative results between centers with these unmatched series of patients. So, using anatomic staging, two of the most widely used would be the Cotton-Myer staging system, which is developed in pediatrics and it's widely utilized and highly reproducible. And it does seem to track with surgical outcomes in adults, as well. What I think is a little bit more precise is the great system developed by Dr. McCaffrey, taking into account the degree of glottic involvement in disease. And these two really are very helpful.

Dr. Alex Gelbard:

There's also been recently a classification system developed by the European Laryngologic Society, which has been validated in adults and peds retrospectively. And this seems to have a lot of promise at precise classification of patients. So, in addition to the anatomic staging that comes about from some of our systems, physiologic staging is also important. And this is, "How well do they breathe? How good is their swallowing function, and how good is their speech and phonation beforehand?"

Dr. Alex Gelbard:

And so, to really do a good job physiologically staging people to give you a mechanism to compare before and after and compare between centers, you really should include documentation of the degree of respiratory impairment, how impaired are their activities of daily living, and explicit lung function testing. Pre and post-operative evaluation of voice should be conducted using some really well established patient reported instruments, and maximum phonation time, and use of phonemes as well

as vowels, and a dynamic voice range profile are really solid vocal parameters to capture pre and post operatively.

Dr. Alex Gelbard:

Swallowing function can be formally assessed with a combination of directed questions, weight loss, aspiration, recurrent pneumonias, along with patient reported outcome tools and instrumental exams. So, a modified barium swallow, or endoscopic evaluations of swallowing function. And then, we also use some laboratory studies to help provide some physiologic stage and where we found red cell distribution width gives you some objective measure of intrinsic host inflammation, and red cell distribution width values above normal, so anything above about 12.5 seem to predict worse outcome with open reconstructions. And we factor these into discussions about treatment options.

Dr. Linda Yin:

And this is something that always confuses me quite a bit, is there any role for pH impedance testing as part of the workup?

Dr. Alex Gelbard:

So, I think the role of gastroesophageal reflux disease is important one in the airway, different centers approach this differently. Some centers will only evaluate patients who report symptoms with more invasive objective physiologic tests, and other centers will subject everybody to the testing to help understand the contribution of disease,

Dr. Alex Gelbard:

and I don't think there's a right or wrong answer. It does seem that controlling gastroesophageal reflux disease when it's present is important to help in mucosal healing after endoscopic or open reconstructions.

Dr. Linda Yin:

And we talked about the importance of a good, flexible laryngoscopy exam in clinic, but is there any role also for direct laryngoscopy under anesthesia in order to work up LTS?

Dr. Alex Gelbard:

Yeah, I think any patient that comes to see me, that's interested in their options to help take care of disease, we include an operative direct laryngoscopy and bronchoscopy. This initial encounter offers an opportunity for refinements in anatomic staging, where you really can assess cricoarytenoid joint mobility. You can really assess the length and the characteristics of the mucosal scar. And I think one of the most important things it lets you do is help delineate the contribution of mucosal or framework pathology to the disease.

Dr. Alex Gelbard:

It gives you the extent of healthy distal trachea that you may have to work with in reconstruction, but it also gives you an assessment of the patient physiology in a controlled setting. What's the ease of their laryngeal exposure, and what's their pulmonary reserve like. Most importantly, for patients that offers opportunity to provide endoscopic treatment aimed at restoring their airway caliber, and that'll improve

their symptoms. So, I think for all the patients that come to see us, an operative endoscopy is part of the workup as we try to delineate what their surgical options may be.

Dr. Linda Yin:

Okay. I think now is a good time to backtrack into pathophysiology. So, let's dig a little deeper now. I understand there are different forms of LTS, and the understanding of the different etiologies can kind of influence our treatment decisions. So, this is kind of a loaded question, I know, but what is the disease process behind LTS?

Dr. Alex Gelbard:

Well, I think all LTS is characterized by a mucosal fibrosis and fibrosis is composed of sustained inflammatory cell activation infiltration, along with an excessive accumulation of the fibrous connective tissue components of the extracellular matrix. So, both matrix and inflammation are present, and host inflammation can also progress to involve the cartilaginous airway framework to render structural loss in the integrity of the airway.

Dr. Alex Gelbard:

So, the different types of diseases probably have different mechanisms of host inflammation and may have different degrees of dysfunction in the fibrous remodeling process. But the basic concept that's universal to all of them is fibrosis. And that's a pretty established concept in a lot of other organ systems.

Dr. Linda Yin:

So, kind of along those lines, what are you seeing on a slide in terms of histopathology?

Dr. Alex Gelbard:

So, histologically, we'll see subepithelial inflammation with a lymphocytic infiltrate, and you'll see fibrosis of the lamina propria with excess collagen. Sometimes you'll see granulation tissue, and sometimes you'll see neovascularization. That pathology and histology are not superb at differentiating the different etiologies of disease. They do help you understand whether there's active inflammation going on, but their contribution to differentiating the subtype of LTS is still, at this point, pretty limited.

Dr. Linda Yin:

Gotcha. Let's go deeper now, into the different etiology. So, can you talk a little bit about iatrogenic LTS?

Dr. Alex Gelbard:

So, in iatrogenic LTS, the cascade of inflammation and fibrosis is set off by a known cause. Intubation injuries occur when an endotracheal tube is placed in the posterior glottis, and the vector forces the tongue base drive the endotracheal tube back into the larynx, creating a mucosal pressure injury at the level of the larynx. Additionally, an overinflated endotracheal tube cuff can injure the mucosa in the sub glottis or proximal trachea, leading to expose cartilage chondritis and structural loss. Animal models demonstrate that after an initial injury, the airway mucosa undergoes a rapid infiltration of inflammatory cells, followed by neovascularization and subsequent progression to mature fibrosis.

Dr. Linda Yin:

And how bout auto-immune laryngotracheal stenosis, what's happening here?

Dr. Alex Gelbard:

So, that's primarily granulomatosis with polyangiitis, what some people would call Wegener's Granulomatosis. Wagner was a Nazi scientist, and so people have been moving away from using that name. And in GPA about 10 to 20% of patients develop laryngotracheal stenosis. And we don't understand the risk factors that predispose GPA patients to airway stenosis, but it does appear that better systemic disease control translates to reduce exacerbations of their airway disease. So, integration in care plans with a rheumatology colleague is really critical. And when their disease is quiescent, there are several series that show good outcome with more invasive open reconstructions for recalcitrant cases.

Dr. Linda Yin:

And now the hard question, what about idiopathic subglottic stenosis? Do we know of any causes behind those?

Dr. Alex Gelbard:

Yeah, it's another interesting disease that we don't yet fully understand, although we're working hard to try to gain some insight, and it is clear that the disease primarily affects adult Caucasian women, and the really tight demographics of the affected population really provide mechanistic support for the idea that a conserve and consistent biologic process drives a singular disease. But the explanation for why involvement is nearly universally restricted to women or the sub glottis is still a little bit opaque. So, we're spending a lot of energy on trying to investigate the role of genetics, the influence of estrogen, and defining the native host inflammation to try to gain insight into the etiology, the fibrosis, that these patients demonstrate.

Dr. Linda Yin:

Okay. I think now that we have a good understanding of the etiologies, let's talk a little bit about treatment. So, we've done our staging, we've done our workup, and now we're going to talk about treatment options with the patient. So, what are some general principles that we should consider when we're discussing and counseling patients on the different treatment options?

Dr. Alex Gelbard:

Well, first and foremost, we have a lot of conversations about the patient's disease and their goals of care, and the risks involved with the different surgical options available to treat their problem. So, sometimes we decide that an airway prosthesis, so a tracheostomy, a T-tube, or a stoma stent provides the lowest risk option to provide voicing, breathing, and swallowing. And some patients really like that. But for patients who are interested in the life without an airway prosthesis and are adequate surgical candidates, we have a number of options. So, in broad terms, when the disease is limited to the mucosa, it can be effectively addressed with either endoscopic or open techniques. But when problems involve the airway framework, open reconstruction to reestablish durable airway patency is really required.

Dr. Linda Yin:

Now, for patients that really don't want an airway prosthesis and don't want a tracheostomy, what are some endoscopic procedures that we can offer them?

Dr. Alex Gelbard:

Well, in broad categories, the endoscopic approaches include dilation. This is probably the most commonly performed procedure. So, either a balloon, or a rigid bougie is used to expand the stenotic segment of mucosa. There are some complications associated with this, although they're infrequent and typically minor, but they can include tongue paresthesia from the direct laryngoscopy, along with dental or mucosal injuries. And very rarely, you can have perforation of the trachea or sub glottis from over dilation, but this is pretty uncommon.

Dr. Alex Gelbard:

Another approach that has really become increasingly relevant over the last five years is endoscopic resection. And in this, a CO2 laser is used to endoscopically excise a quadrant of the stenotic airway, taking care not to make a circumferential injury to promote contracture, but just removing discrete quadrants. And then, patients afterwards are maintained on a longterm medical regime, including an inhaled steroid, a proton pump inhibitor, and Bactrim. This approach, similar to the dilation, include complications like tongue parasthesias, dental injuries, and because it has a intensive medical regime associated with it, there are some adverse side effects from the medical regime, as well.

Dr. Linda Yin:

Yeah. You talked a little bit about those systemic medicines that can be given, but how about local or topical therapies during endoscopic procedures? Is there any role for those?

Dr. Alex Gelbard:

So, many people use intralesional steroids and it certainly doesn't seem to hurt at the time of the OR, and it would make a lot of sense given the histology demonstrating a immune cell infiltrate for the steroids to help. Mitomycin C's another drug that's used. There's certainly mixed evidence on its efficacy and probably all in all, given the risk profile as well as the drug handling, given that it's a chemotherapeutic agent, this is used a lot less frequently nowadays than it used to be.

Dr. Alex Gelbard:

It doesn't seem like there's really strong evidence that this dramatically alters disease course. Now, other people have tried injecting corticosteroids directly in the subglottic scar in the clinic. And there's some evidence that this may be promising, and we're still waiting for bigger studies to confirm the efficacy of this approach, as well.

Dr. Linda Yin:

Moving into the more invasive procedures now. So, outside of the endoscopic procedures, are there any open airway procedures we can offer patients?

Dr. Alex Gelbard:

Well, when I was going through my training, I always felt like this was confusing. There was a bunch of different things described, and it was hard to find some sort of theoretical framework to link them all together. And the details of the varying techniques are pretty extensive. But really for the purpose of this talk, problems of glottic mobility or laryngeal stenosis really need to be recognized and addressed as a component of therapy.

Dr. Alex Gelbard:

When you're talking about enlarging the tubular sub glottis or trachea, there's really three primary mechanisms to do that. So, you can expand the airway via a combination of anterior and posterior grafting, what we would term a laryngotracheal reconstruction or LTR. You can resect the airway, ie. remove deficient airway structure and so healthy proximal and to the distal airway, this includes a tracheal resection or a cricotracheal resection.

Dr. Alex Gelbard:

The other thing you can do, is you can slide the airway. So, you divide the deficient airway structure, but rather than remove it, you splay it apart at the proximal distal segments and on lay the two segments to widen that segment of the airway. So, expanding, resecting, or sliding the airway are the three main open approaches to address subglottic and tracheal disease.

Dr. Linda Yin:

Now, I know it's kind of difficult to talk about complications when we're talking about such a wide variety of procedures. But in general, with open procedures, more so than endoscopic procedures, what are some complications that are unique?

Dr. Alex Gelbard:

Sure. Well, there are some risks specific to these types of procedures. So, you can have an anastomotic complications, and this can be granulation tissue, restenosis of the suture line, and anastomotic dehiscence, you can have tracheoinnominate fistula and tracheoesophageal fistula. You can also have a whole range of non-anastomotic complications.

Dr. Linda Yin:

And after you've performed a procedure on patients, whether that's endoscopic or open, how do you typically follow up or perform surveillance on them?

Dr. Alex Gelbard:

So, my goals in follow up surveillance are to monitor the efficacy of my therapies and detect recurrent disease early to avoid emergency compensation, and also to really ensure phonation and swallowing are optimized postoperatively.

Dr. Linda Yin:

Now, I understand that sometimes we'll tell patients to track their own progress or their own disease at home with a peak flow monitor. Can you explain this a little bit, and what we're measuring here?

Dr. Alex Gelbard:

Yeah. So, peak expository flow rate measures the highest flow during exploration, and it's a sensitive and specific measure of disease progression in the sub glottis. And it can be measured at home by patients using a handheld meter that's really, pretty inexpensive, the same type of meter that we'd give to patients with asthma. And people have found that it's very helpful to track their disease progression on a day to day basis. It offers some degree of remote monitoring, as well as something along the lines of biofeedback for patients who are wondering about how their air was doing.

Dr. Linda Yin:

Okay. I think those were all the questions that I had on LTS. Is there anything else that we haven't covered that you think would be important to cover, or any other point that you want to emphasize for the listener?

Dr. Alex Gelbard:

No, I think that's a great start. Anybody who spends a lot of time taking care of these patients realizes that there's still a lot of questions that are left unanswered, but I think the questions you asked are great start to deeper understanding of the disease.

Dr. Linda Yin:

Awesome. Well, thank you so much for being here today.

Dr. Alex Gelbard:

Thanks for asking me.

Dr. Linda Yin:

Okay. Moving on to the summary section, now. Laryngotracheal Stenosis or LTS is a collection of diseases that share a common structural problem, which is luminal narrowing of the upper airway that can involve the glottis, the subglottis, or the trachea. LTS can be divided into several different subtypes based on the etiology. Iatrogenic LTS is by far the most common subtype, and this is related to scar formation after endotracheal intubation.

Dr. Linda Yin:

The next most common is going to be idiopathic subglottic stenosis and autoimmune disease, which each makes up about 50% of cases. Idiopathic disease, of course, has no known cause. And in terms of autoimmune disease GPA, or granulomatosis with polyangiitis, makes up the most of this disease. Patients with LTS typically present with dyspnea, this is the most common symptom, although hoarseness and dysphasia even are possible.

Dr. Linda Yin:

The diagnosis and categorization of LTS can be accomplished with a careful history, physical exam, sometimes laboratory studies and pulmonary function testing, and also radiographic studies, most commonly a non-contrast CT. Pulmonary function testing and LTS shows a fixed extra thoracic obstructive pattern, which can distinguish it from other intrathoracic obstructive diseases like asthma, and also intraparenchymal pulmonary disease.

Dr. Linda Yin:

LTS can be staged either with an anatomic systems such as the Cotton-Myer scale or the McCaffery system, or it can be stage physiologically. Idiopathic subglottic stenosis has a very homogeneous patient population, as it typically presents in otherwise healthy middle-aged Caucasian women. Iatrogenic LTS is typically associated with a history of intubation, and risk factors for this can include traumatic intubation, prolonged intubation, or intubation with a large tube or high cuff pressure. In GPA about 10, 20% of patients will develop LTS. And the local disease severity does appear to be related to the overall

systemic disease control. Histologically, LTS is characterized by subepithelial inflammation, which has a lymphocytic infiltrate as well as fibrosis of the lamina propria with access of collagen deposition.

Dr. Linda Yin:

Treatment in LTS is tailored to the individual and their specific needs. The main question is, does a patient want a longterm airway prosthesis such as a T-tube or a trache, or would they rather prefer a more definitive surgical therapy to achieve a more sustainable natural airway? Surgery can consist of endoscopic approaches or open reception, and endoscopic treatment can include either dilatation with a balloon or CO2 assisted endoscopic laser resection. Open procedures can be categorized into a few main categories, there are surgeries that can expand the airway using a combination of grafts. There are those that resect airway scar, and then anastomose the different segments together, and those that slide the airway to allow it to widen.

Dr. Linda Yin:

Okay, we're moving on to the question section, now. So, for this part, I'll be asking some questions that highlight key points from the talk, and then I'll give a brief pause before providing the answer. Our first question is LTS has several different subtypes. What is the most common subtype in terms of epidemiology? The most common subtype of LTS by far is going to be iatrogenic LTS. This accounts for up to 70% of the disease when looking at large series from airway centers around the world.

Dr. Linda Yin:

Next question, what laboratory studies can be helpful as part of the workup for LTS? Lab studies can be helpful to rule in or rule out autoimmune disease, specifically GPA. So, the test to get will be cANCA testing, specifically cANCA antibodies, which are commonly targeted against PR3, or MPO and GPA.

Dr. Linda Yin:

What type of pattern is seen on pulmonary function testing for LTS? LTS has a fixed extra thoracic obstructive pattern on PFT, as seen in the flow volume loops. This means that volumes are diminished both during the inspiratory and expiratory phases of the disease, and this distinguishes it from intrathoracic variable obstruction or other interproximal pulmonary diseases.

Dr. Linda Yin:

What does the pathology look like for LTS? LTS is a disease of mucosal fibrosis that can have an active inflammation component. Histologically, you can see subepithelial inflammation with lymphocytic infiltrate, as well as fibrosis of the lamina propria.

Dr. Linda Yin:

And last question, what are the most common types of endoscopic interventions available for patients with LTS? Airway dilation is probably the most common performed procedure overall for LTS that involves the subglottis or trachea. And here, a balloon more often, or otherwise they're rigid bougie, can be used to expand the stenotic segment. Another common procedure that is emerging in the last few years is endoscopic laser resection, and here, a CO2 laser is used to resect quadrants of the stenotic airway. That's our show. Thanks for listening. And come back next time.