Dr. John Marinelli:

Hello, and welcome back for another episode of ENT in a Nutshell, my name is John Marinelli, and today we're going to be talking about cricopharyngeal muscle dysfunction with laryngologist Dr. Greg Dion. Dr. Dion, thanks so much for being here today.

Dr. Greg Dion:

John. Thanks a lot. I'm looking forward to contributing and being part of ENT in a Nutshell.

Dr. John Marinelli:

Great. Well, we've got a lot of exciting material to cover today, but we'll just start in our usual fashion with disease presentation. So Dr. Dion, how do patients with CP muscle dysfunction typically present?

Dr. Greg Dion:

Whoa, I guess you're starting with a zinger here. That's a little bit tricky, John. Unlike thyroid surgery or tonsillectomy patients often present with a really vague and nonspecific findings. So you might see someone who's coming into your clinic with a cough or trouble swallowing or just even throat clearing. And so you can envision how that overlaps with a lot of the other things and diseases that we deal with as an otolaryngologist. So you got to find a way to break that down. I think one really nice way to do that is to look at the epidemiology of, "What's the age of the patient?" We know that cricopharyngeal muscle dysfunction is more common in the aged population, so you're less likely to see that in a young person. So that can be helpful. You also can look at where these patients are coming from.

Dr. Greg Dion:

Are they coming from a gastroenterologist? Are they coming from a speech language pathologist, a neurologist or a medicine doctor? That gives you some mindset of where they're coming from. So someone from GI is more likely to have a cricopharyngeal muscle dysfunction problem than say someone that's just coming from general medicine with a cough. Some of the symptoms though that are guiding hints would be globus, meaning the sense that there's something in the back of the throat, a frequent throat clearing and that's also associated with the sense that they can't get some food down as in, "Oh doc, I take a drink of water and there's just something there it doesn't seem to all go down, I got to swallow two times." So you could talk about a double swallow and I mentioned, they may have trouble swallowing, perhaps solids or liquids or some combination thereof.

Dr. Greg Dion:

Then there's some obvious things that can be hints such as aspiration pneumonia history, meaning that you're already thinking that they probably have some sort of esophageal or aortoesophageal dysfunction. So I realized that that's a hodgepodge of things but when we start thinking about cricopharyngeal muscle dysfunction, it's really a broad topic. And so you start thinking about things that would happen in the pharynx when you're not clearing stuff such as like throat clearing, cough, aspiration.

Dr. John Marinelli:

Yeah. No, that's helpful. Before transitioning to pathophysiology, I think this topic really lends itself well to talking about some normal anatomy and normal physiology. Could we first just review the anatomy of the UES, the upper esophageal sphincter?

Dr. Greg Dion:

Yeah, absolutely, John. So even starting with our naming conventions, we get ourselves into conflicting stories depending upon what you read. So what frequently is discussed as the UES or upper esophageal sphincter is often referred to as well as the pharyngoesophageal segment. When we look into literature that the beauty of that PES or pharyngoesophageal segment naming convention, is it really tells you about that area that you're looking at, not just one muscle. Because when you think about it, if you look at the UES or the PES, really what it's comprised of is the inferior constrictors from your pharynx, the muscle fibers of the cricopharyngeal muscle itself, as well as the proximal esophageal muscle fibers, all coming together. So not to go too far down a rabbit hole. So I'm never one to resist a good rabbit hole.

Dr. Greg Dion:

You can look at the cricopharyngeal muscle itself as a C-shaped muscle. So it's coming off the back portion of the cricoid. Interestingly enough, there's no midline RFA as it comes around the posterior part of the pharynx. So there's no midline RFA associated with the cricopharyngeal muscle. It's approximately one to two centimeters in height. If we're looking on a person standing up. So you're talking about one to two centimeters in height made predominantly of type one, slow twitch fibers. And so you can think about ... that makes sense. We're not talking about, your bicep as you're out there doing curls to look good for people in the gym. The slow twitch fibers reflect the normal function of what you'd find in the cricopharyngeal and other smooth muscles.

Dr. Greg Dion:

Then interestingly enough it reacts different than a lot of other muscles. So the maximal tension in the CP muscle has been founded about 1.7 times its basal length predominantly with a high level of connective tissue. And so this is really similar to what you see in the heart, meaning as the heart fills the contractile strength increases. And so similarly, as the ... you distend the PES or the cricopharyngeal muscle. its contractile ability strengthens too. And then when we talk about anatomy, we always include innovation patterns. And so the innovation of the cricopharyngeal muscles variable. So if you read the literature, you'll find reliable findings of recurrent laryngeal nerve, superior laryngeal nerve, as well as cranial nerve nine innervation patterns. So again, as with the presenting symptoms, there's a lot of nonspecific issues when you're talking about the PES.

Dr. Greg Dion:

And then when you're looking for, where does that lie anatomically in the back of the throat? It lies just anterior to the buccopharyngeal fascia, which obviously is just anterior to the retropharyngeal space, which posteriorly you run into the ALAR fascia, then the dangerous space and the prevertebral fascia. So it lays in those fascial planes that we as otolaryngologists find so important for tracking infections and anatomical dissection. So those are the kinds of things that I think are really important to understand is the space we're working on how it's innervated and what's in that general area.

Dr. John Marinelli:

And what about normal swallow physiology? How does that work?

Dr. Greg Dion:

That's another good one, John. We could spend a whole hour talking just about swallow physiology, but for the purposes of understanding the cricopharyngeal muscle function, we can think of a normal swallow starting as the oral preparatory phase. So in that phase, you're preparing that bolus you're

chewing it, you're grinding at the ... enzymes are starting to work in the saliva. You're making that bolus smooth so it can then be transferred into the pharynx. And then the pharynx itself, squeezes. As the pharynx squeezes, the larynx moves anteriorly, superiorly opening the pharyngoesophageal segment, the weight of the bolus then moves back, rests upon the epiglottitis, causing the epiglottitis to flop over the airway. This then passes into the esophagus. Then the pharynx relaxes the larynx descends and moves back posteriorly. And now you're in the esophageal phase, the swallow. So that's a really broad strokes overview of what happens in the swallow, but it gives you a sense of how dysfunctional long that pathway could create a problem.

Dr. John Marinelli:

Yeah, I agree. I think, it's really helpful. And what about specifically with CP dysfunction? How are patients normal swallow physiology affected by this disease?

Dr. Greg Dion:

Yeah, sure. So looking back at what we were just talking about, you could imagine, where does the CP fall in that pathway? Well, the cricopharyngeal muscle has to be able to open meaning, be stretched, open passively, and actively allowing things to go through. So any dysfunction that affects that would be problematic. So say for example, you have a hypertonic disorder, so maybe there's radiation induced fibrosis of the muscle, which prevents it from stretching out. So you have fibrosis problems. You could have some kind of muscle dysfunction muscle dystrophy, you get hyper-tonicity with muscle dysfunction, and now it's so tight that you can't get it to loosen up. You might also have a disorder in which it's flacid. So if you have a neuromuscular dysfunction in what you have a flacid UVS, you could be predisposed to regurgitation, aerophagia meaning swallowing air, subsequent burping. I can also be affected by a neurologic insult.

Dr. Greg Dion:

So for example, perhaps you had a stroke or ALS and you have neurologic issues. So anything that would affect the nerve flow. So stroke, nerve injury, et cetera, is going to alter the timing and the way that the CP muscle is going to function in conjunction with all of those other swallowing physiology. So you could imagine if for whatever reason your muscle was slow to react, you'd now have a pharynx squeezing. So your pharyngeal squeeze is pushing against a tight pharyngoesophageal segment or a cricopharyngeal muscle in which case, the bolus isn't easily passing through. And the things that you would end up with would be like retained bolus, et cetera. And so these things all contribute together and could create ... you could see how any dysfunction there would predispose somebody for having an interrupted swallowing physiology.

Dr. John Marinelli:

Yeah. And you started to get into this already, but is there anything else on your differential for CP muscle dysfunction that you're thinking of that could be overlapping in terms of their presentation and whatnot?

Dr. Greg Dion:

Yeah. This was one of those things we look back and we laugh about, "Well, what does the patient look like that shows up with CP muscle dysfunction?" Well, that's a vague statement. That's one of those picture covers for diversity on the magazine where you've got a little bit of everything. Well, so we have to make sure we rule out all of those other things that could be contributing. So we get down at the end

of the day to just the cricopharyngeal muscle. So for example, you might have a weakness leading to a diverticulum. So a Zenker's diverticulum, a Killian Jamison's diverticulum. You can have other things that create dysphasia. So an esophageal web those can be a little tricky to diagnose, you find an esophageal web that could cause trouble.

Dr. Greg Dion:

You can have eosinophilic esophagitis. That's another one of those that's really hard to diagnose. And so we're quick to forget about it or diagnose it without fully going through a real careful workup. You could have a motility disorder. So when we think about, does that patient have secondary cricopharyngeal muscle dysfunction? And this is one of my big pet-peeves, is we say, "Oh, Hey, there's a CP issue." You're going through your diagnosis, you're talking to someone and you're like, "Oh yeah." But at the end of the day, you got to make sure that that CP dysfunction is not secondary to another problem such as motility. You could have lower esophageal sphincter achalasia, and again, causing a secondary cricopharyngeal muscle dysfunction.

Dr. Greg Dion:

In addition, a careful history would elucidate other potential reasons to have these swallowing complaints. It could be an autoimmune disease where you get autoimmune esophageal dysfunction, and then there's some simple things that we tend to see a lot and frequently in otolaryngology practice that really need to be worked up. "Now, has this person been worked up for seasonal allergies, food allergies, postnasal drip, sinus disease?" And so working through those things as you rule those out, then you're starting to say, "Yeah, the findings that I'm seeing on physical exam testing, et cetera, that are consistent with cricopharyngeal muscle dysfunction, aren't actually secondary to, or because of another reason."

Dr. John Marinelli:

Yeah. And just building off of that discussion. So if you're in clinic and you're seeing someone that you suspect might have CP dysfunction, what does your workup look like?

Dr. Greg Dion:

Yeah. Great question. Again, as much as I'd love to be Rachel Ray here and give you a recipe, it's a little bit of how that patient presents. So, I need to rule out those other disorders, we look back and we say, "Yep, no Zenker's." So I have to figure out how do I do that? Well, so that patients now in my clinic, so I get to rely on the good old fashioned history and physical. So some of the things in the history are going to tell me a lot of information. So does that patient have a history of autoimmune disease? Do they have a history of say PH proven gastroesophageal reflux disease? So those things are going to help me understand that. And then there's, the next level of exam. So we can do some simple things like a clinical swallowing evaluation.

Dr. Greg Dion:

And I think this is really under appreciated. We're ready. We've got a scope in our hands and we're ready just to go after that patient with the scope. But the reality is sitting there and watching them talk and maybe take a sip of water and you'll notice do they drink that water from their water bottle in one sip and one swallow, or do they take a sip from the water bottle? And then you see them struggle to propel that water. And then they have what appears to be a hard swallow or even a second swallow to get it down. And so those things help. They might swallow and start coughing. That's obviously not ideal

in your clinic, but this is important to notice. Depending on your clinical setup, you might watch that patient come down the hall to the clinic room.

Dr. Greg Dion:

Now do they have a shuffling gait? Is it clearly a patient with Parkinson's? We see this all the time, these patients haven't been anywhere else, these are things that we as physicians need to be looking at. Do they have something wrong with their voicing pattern? That's going to lead you to believe there could be a neurologic issue contributing to that. So those are some of the easy things before you really get too involved. When you start thinking about what are the most useful tests, I would start to break them down into ones that you can do right there in clinic. So flexible enteroscopy plays a really important role in understanding what's going on in cricopharyngeal muscle dysfunction. So you want to take your time as you get the scope back there and take an opportunity to look at the overall anatomic function.

Dr. Greg Dion:

You need to make sure the vocal folds move properly, because if you have a unilateral paresis or paralysis, that could be predisposing you to aspiration and the cough, which you originally thought could be CP dysfunction. One of the great things you can have patient do is try to blow out against area with their mouth closed, where I'd say, puff their cheek out ... I always say, "Puff your cheeks out like a bull frog." And you can then up those piriform sinuses. You can get the camera in there and see is there pulling of secretions? Is there, can you see, today's pills and the Zenker's diverticulum and the post trapezoid space, that's possible to do with that maneuver in clinic really allows you to see what's going on and really be thoughtful with that exam. I take a moment here and say, one of the things you want to be careful of when you do these exams is you don't want to drown the patient in your nasal spray.

Dr. Greg Dion:

So all too often a technician or a really eager clinician or resident, just sprays multiple CCs of [inaudible 00:15:34] and Afrin and the patient is coughing everywhere, gagging certainly not ideal in the COVID pandemic, but the other problem is looking back and what's been done. You can see that if all of that stuff ends up ... and that [inaudible 00:15:50] ends up in the pharynx, you can end up with pulling in secretions that's distracting. So, this is really important to look at. And I think that the final thing in clinic you could consider is FEES, a functional endoscopic evaluation of swallow. So traditionally we think of a FEES as something just a speech pathologist does, but in reality, this can provide us a lot of information. Remember that depending upon your practice setting, sending that patient out for a different swallow study, A, might cost additional money or not be approved, it's a different visit on a different day.

Dr. Greg Dion:

So depending upon your clinic setup, that's the other good or bad. And so this is something you could do, provided again, you haven't gone through and sprayed too much spray in there, and you're going to give them different textures and consistencies and volumes of food, and see, are you seeing retained bolus? Are you seeing stuff pulling in the secretions? Are you seeing penetration or even aspiration in your exam, and then you're really going to make a thoughtful move as to what you do next. So I'd say, from a clinic standpoint, after you're done doing your complete exam and feeling the neck for any restriction, if that patient has had ... say they've had radiation in the past, and you can feel that they don't get high laryngeal elevation when they swallow you know ... you're thinking, "Okay. Yeah, I get it. This is probably leading to where we're having trouble."

Dr. Greg Dion:

So I'd say that's really what you're thinking when you're in clinic. And certainly not all of your patients need a FEES, but it's worth considering. So then you have to decide like, "What else can I do for workup?" Well, that could lead to us down a number of pathways, but just as a brief overview from probably least likely to most likely, I guess you could do quick pharyngeal electromyography. Well, that's useful mainly for research, but it's technically very challenging because you got to put that needle into the CP muscle, which is moving in the neck and the patient with ... depending upon your part of the country, varying neck circumferences which could be quite challenging. And it's hard to really get a reliable reading from that.

Dr. Greg Dion:

And so not commonly done. A little bit more common, but still not the most common, and yet would be high resolution manometry. I promise I won't get too excited here, but this is a fascinating way for us to understand what's going on in the esophagus itself. We're going to learn with HRM or high resolution manometry exactly how that bolus propels all the way from the entrance to the esophagus. And really if you look at the latest data and work really from the pharynx and we're measuring some pharyngeal manometry all the way down to the stomach. So are we seeing some failed peristalsis, jackhammer esophagus, or potentially elevated LES pressures? I'd hesitate to talk about findings on HRM as far as the UES or PES are concerned, because really at this point, I don't think clinically there's enough data out there to support, making objective measures or movements forward based on what you're seeing on HRM.

Dr. Greg Dion:

But I think as far as rolling out some other dysfunctions, it's really a key thing that as an otolaryngologist, we need to embrace both as a diagnostic means. It's something that we should be considering participating in. And then I'll move into our two related most common studies. One would be your esophagram or your barium swallow. This is where you send the patient down to radiology, and they do a study where the patient drinks, a large volume of high molecular weight barium, which really lets us fill the esophagus and get a good picture. You can see some esophageal wall abnormalities. You'll also see potentially dysmotility. If you look carefully in some patients with eosinophilic esophagitis, you might see some ridging on the edge of the esophagus. And that can be a hint in some cases, but a lot of times you miss the pharyngoesophageal segment.

Dr. Greg Dion:

And of course despite growing data firms of research, for some reason, it seems that we only get a little bit of data saved out of esophagram, which happens to be captured to just six frames per second. Which to me is quite amazing, because there's no way you're watching YouTube at six frames per second. And that's what brings a sense of the video fluoroscopic swallow study. So there's a number of reasons I left this to last and as our like main go to study, this is recorded at 30 frames per second. So you're really able to tease out the nuances of the swallow all the way from that preparatory phase into the esophagus. And in many cases, depending upon your institution, a lot of these will have an esophageal screen or follow through afterwards.

Dr. Greg Dion:

And so in this study, the speech pathologist is giving various bolus sizes of solids, liquids and some mixed consistencies to give some answers as to what's happening in the swallow. And this is where you're

going to see evidence of maybe incomplete pharyngoesophageal segment distension with retained bolus above the PES segment. You can envision in your mind where you would get the proverbial cricopharyngeal bar, CP bar, or depending upon how the speech pathologist might word it as, "A tissue section, pushing off the posterior wall of the esophagus." And so understanding how to interpret that reading is important to say, "Okay, so the video for scopic swallow study is really going to be one of our key studies here." So those are the kinds of things you can order. I think when you're doing a workup, one of the things you can't forget about is the role of endoscopy.

Dr. Greg Dion:

And so we talked a little bit about laryngoscopy and you do that in the clinic with our patients, but at some point in this workup, you need to consider visualization and/or biopsy when needed. And so that would take the form of either transnasal esophagoscope. So in the clinic you have the patient come in, they saw the esophagoscope passed down to the stomach, and then you can really perform a thorough, good evaluation of the entire length of the esophagus to determine, "Are we missing any masses, lesions? Is there trachealization of the esophagus, which is making us think of EOE? Do I see a Schatzki's ring and that I can pay my hat on?" And these are things that will help you understand. A lot of practices may not have a TNA scope in their clinic. And so in those cases, you can send a patient for a formal esophagogastroduodenoscopy often this has been done though.

Dr. Greg Dion:

When you look at these patient histories and take a careful, careful history, you'll notice like, "Oh, I had an EGD two years ago. Here's the report. It looked normal." Well, that's fine because really what you're trying to do with this is rule out the big things like you really don't want to miss a cancer. You don't want to miss an obvious disfunction candida of the esophagus. You don't want to miss any of those things. And then the other thing you can do is if you're planning an intervention and you're down the pathway, like, "Hey, I think I know this is going to be CP dysfunction. I think I can intervene and potentially help this person." It's certainly possible to do ... esophagus can be at the time of your surgery and the patient's asleep already, is not two sedations.

Dr. Greg Dion:

So without having gone too far off the rails here, and when you asked for a typical workup, that's very nonspecific, but you can imagine that you have all these tools at your disposal and the patient's story's going to push you down, one of these pathways. And so, I would say that everybody needs some swallow evaluation, whether it's HRM, esophagram or video swallow, I don't think you can get away without having at least one of those reasonably to make a diagnosis. And I think there needs to be some form of visualization, be it a TNE, EGD or esophagus sphincter in surgery. So while not trying to give you too much information, hopefully it answered your question on how I might work that up.

Dr. John Marinelli:

No, I think that's very helpful. Dr. Dion. I think as we start transitioning to talking about management and whatnot, one question I like to ask is, when you're counseling, these patients that have evidence of cricopharyngeal muscle dysfunction, how do you describe to them the natural history of their disease and what happens if it's left untreated?

Dr. Greg Dion:

Yeah. This is a great question. And this is an area where I think that we still don't totally understand yet. There's been this talk for a long time like, "Okay, well [inaudible 00:24:11] two centimeters Zenker's [inaudible 00:24:12] was just going to get bigger or is it?" We don't have a series of patients just walking around with Zenker's for 50 years that we're able to follow, mainly because we don't find them until they're very old and then they tend not to live 50 years. But that's the thing, we don't really fully understand it. So what we can do is be honest with the patients. We're that way with all these disorders, we can say, "Listen, here's what we know. We've done this workup. We know it's not cancer, because we worked all of that up and we're very confident in our diagnosis." So we can work it up and say, "Hey, this is what we think it is."

Dr. Greg Dion:

And it may never change. It may not be a problem. Remember ... I didn't really mention it, but if we looked at esophgrams or videofluoros of people and in the elderly population and well, over 10% have a "CP bar". They may not have symptoms at all, but they have that. So just because you see something doesn't mean that's the problem, number one. But it may never change. And so a reasonable thing might be, "Hey, let's watch this and see if it does change. The symptoms, aren't bad. Let's see what happens." Is an opportunity. And sometimes it doesn't and just that cognitive understanding of what's going on, that it's not cancer, lets that fall to the back of their mind. I tell people like, "You don't realize you're breathing, unless I ask you to think about taking a breath in and out, you've been breathing this whole time."

Dr. Greg Dion:

Hopefully if you're not like asleep or crash your car, listening to me or something, but if that's the case and you're awake, you're breathing, you're just not thinking about it. So if we let the patient understand, there's nothing bad and they've forgot about it. That's really helpful. It's possible of course, that it could get worse. And when we talk about patients that are ... I call them the "late rad patient". So a patient had radiation and now fibrosis is setting in and they used to go out and toss back a steak, and now they're doing milkshake and losing some weight and some say, "Well, that could be your course ... your disease course." We know that fibrosis could worsen with time if there's no intervention. And then one of the things that can happen with that is aspiration pneumonia. So it's really hard to tell a patient like this is what happens, but I think education and collective decision making really helps you there.

Dr. John Marinelli:

Yeah. In the scenario of a symptomatic patient who has evidence of a large obstructing bar and Videofluoroscopic Swallowing Study, what are the different management options for these patients?

Dr. Greg Dion:

Yeah. So in this case, I'm not sure exactly how much per say size matters, because we don't have enough data yet, we'll look at and say like, "Oh, that huge bar." I think the collective findings of the patient are what we really want to think about. So there's an obvious cord extension of the pharyngoesophageal segment. Not a lot of stuff is getting through those routine bolus. The patient is coughing after they eat. There's concern. They've been admitted for one aspiration pneumonia. Yeah, then we needed to do something. So probably the option of observation and reassurance, probably not the best, because this person's clearly having symptoms in trouble. If we were unable to determine if that person had any history of gastroesophageal reflux, a PH study to assess the actual presence of reflux, which is no one's

favorite cup of tea, because we'd want to get a PH with impedance, meaning that the patient be walking around for 24 hours, a little wire out of their nose.

Dr. Greg Dion:

And obviously that's not everyone's first line of fun. So you could give them a trial reflux meds and say, "Well, maybe, this is related to underlying reflux meds." Because interestingly enough, we know that if we expose the lower part of your esophagus to acid, so if I held you down and put acid on your lower esophageal sphincter, your upper esophageal sphincter will in turn contract. Even if itself is not exposed to reflux. So as a result, we want to make sure that we didn't miss a reflux, because that's easy if you have reflux and we treat it and your upper esophageal symptoms get better, that's fantastic. So those are some easy things. You also believe it or not, there is a speech component here. You can do some biofeedback swallowing exercises where you show the patient, they're able to swallow on a flexible laryngoscopy.

Dr. Greg Dion:

And then they notice and they're swallowing and they do well. I mean, you do Mendelsohn's Maneuver. And then this is like you swallow and then you keep that he keeps the larynx elevated, basically a supraglottic swallow to build up the muscle. And that's really truthfully the Mendelsohn Maneuver is the only maneuver that's been shown from a swallowing exercise standpoint to have some impact on isolated pharyngoesophageal segment dysfunction. As we move down the line, we talk about CP or a dilation versus chemodenervation. So this is a little complicated because years ago we thought like, "Well, if it's a tight, we can inject Botox." Well, that's going to work if we're talking about that hypertonic state. Like this hypertonic, and it's not too large. So if it's just a tight hypertonic, pharyngoesophageal segment, we can do chemodenervation of the cricopharyngeal muscle.

Dr. Greg Dion:

And in, so doing, we decrease the tonicity and you can get fluids through. However, interestingly enough, if this is what we had referred to as a big CP bar, you now potentially have created a large flacid obstruction to the esophagus. And in some cases, worsening their swallow. So you got to be thoughtful in terms of where does Botox roll in there? I'd say a lot of people now maybe about 20% of the time, will do some Botox. The dilation is great, because the dilation gives you a variety of options. You could do a traditional dilation where the patient goes to sleep and you take either a bougie or a balloon and you dilate the CP under direct visualization. And in, so doing, the patient wakes up, goes home, eats food and see if they get better, and for how long. Alternatively, a really great way to do this, particularly in patients of whom are not great candidates for general anesthesia, you can go to a procedural suite or an OR.

Dr. Greg Dion:

Though it's been reported in the clinic. I tend to shy away from that a little bit because of discomfort and in a procedure room or the OR. You can give the patient just a little bit of say, Alfentanil take a transnasal esophagus scope, put it through the nose, have them swallow it. So now you're in the esophagus, the patient's looking at you, you're all high fiving slide a thin guidewire through the esophagus scope, pull it out and then slide the balloon over the guidewire and follow it in with the esophagus scope. And you can do an awake transnasal balloon dilation. And in fact as I do not infrequently, you can go in with two balloons, either through the same nostril or other nostril. And so do

a double balloon dilation. If you think about it, that the best study on the actual shape of that open UES was done in [inaudible 00:31:11] or sheep.

Dr. Greg Dion:

And they found that it's a kidney bean shaped. And so you can envision the two balloons that way work well. If you're doing a transnasal esophageal dilation, then you're stuck with using balloons. If you're doing it under direct, you can use other balloons and bougies. And so, while there's no head to head study, there's probably pluses and minuses. There's not a onetime use for a bougie, but there's some thought that the shearing forces versus the lateral fortunes are different between boogies and balloons yet that hasn't been fully teased out in study. So, say you've moved past that though. So you've seen this patient, you tried observation. They didn't like it. They didn't do well, reflux meds didn't help. They weren't really a great candidate for whatever reason or didn't perform well with Mendelssohn's Maneuver.

Dr. Greg Dion:

And, you did a dilation, the patient came back, brought your Christmas cookies was awesome. Everybody's happy, they were doing much better. And then of course with time they got worse. And now they're like, "Man, I got to do something. So the end of the line, so to speak. And when you're really in there to longterm solve, this is the cricopharyngeal myotomy. So the cricopharyngeal myotomy is where we're actually ... I mean, right in the word, we're cutting that muscle. We're trimming the cricopharyngeal muscle. So what are your options there? Well, traditionally we look back a couple of decades and the option was, it was going to be a small incision in the neck and you're going to trim really three to five centimeters, believe it or not the inferior constrictors through the cricopharyngeal muscle. And then down into the cervical esophageal fibers with a bougie in place in the esophagus so that you can feel.

Dr. Greg Dion:

And it's really an elegant procedure because you're in there, it's a small incision in the neck you can see and you feel the bougie underneath it and you watch those muscle fibers cut and then open up and release the pharynx. And so there's really a little bit of elegance to that. But as with many things, everything is gone and endoscopic. And so what's the beauty of endoscopic? Well, it's really cool because you're using lasers. Lasers are fun. Everyone likes lasers, but also that patient does have an incision on their neck. And so it's really come to the forefront of treatment and it doesn't take that long and you can really get outcomes with this. So endoscopically, you go suspend the patient, look at the ... identify the cricopharyngeal muscle. And then you have a couple of options on what laser to use. And that's probably beyond the nuances of the talk here, but most often would be your carbon dioxide laser, which is 10,600 nanometers.

Dr. Greg Dion:

And so you're using that to cut through the muscle, you're going to cut through the mucosa then through the muscle, you're going to see the buccopharyngeal fascia. Remember you don't want to cut through the buccopharyngeal fascia, then you're going to end up in the retropharyngeal space, which is not the plan. So you're going to stay, [inaudible 00:34:00], you're going to see that nice yellow shin, the buccopharyngeal fascia. The other thing that you can use is the thulium laser and that's 2,013 nanometers and the thulium laser is really, really effective, because it comes on ... It's pretty thin. You

can get it on a long extension. You work with that. Nowadays the CO2 laser also can be other line of sight with a micro manipulator or you can use it under a fiber.

Dr. Greg Dion:

So that'll help you, depending upon your angle and your exposure. I would remiss to say that the latest fad which I do actually find useful is going back after you've done the myotomy or even in some cases, a myectomy. And by a myectomy, I mean taking a little piece of that muscle out, the cricopharyngeal muscle and sewing it up and sewing the mucosa back together afterwards. I think that's a little bit less discomforting for patients afterwards. And I suppose how you treat patients afterwards will vary. I guess if we pulled all of my colleagues in laryngology, you'll hear a variety of responses. Some people leave an NG tube in, or a Dobhoff tube in some people feed them right away. I tend to fall somewhere in the middle. I think that, I'll give a patient a clear liquid diet that same night.

Dr. Greg Dion:

So they'll have clear liquids that night till the next morning. And so long as they're improving and doing well, they have a really a soft food diet and able to leave the next day. So, traditionally we used to keep people and we thought about doing the Zenker's and big myotomes in the hospital for a long time, but really now it's the same thing. If they do well overnight, you can give them some [inaudible 00:35:30] and send them home. So really that's the progression through all of the options you would have currently for treatment of these disorders. I would say that they're not all perfect for everyone. If a patient has had late radiation changes, diving in for my anatomy might not be your best option, seeing where you can get with a dilation or even, I call them a pack of three or serial dilations at predetermined intervals, say, six weeks apart. You're going to see that if you can break that fibrosis over time and improve that swallow. So, this is where there are these fine nuances and arts that we're sorting out with science to get better.

Dr. John Marinelli:

And moving on to complications. Maybe just talking first about the conservative interventions, like CP dilation or Botox even what complications do you have to watch out for?

Dr. Greg Dion:

Yeah, so really if we start at the very, very beginning, observation, really, you're going to need to make sure that the patient's not going to be at risk for an aspiration pneumonia. So observation, really that's your thing is are they worsening? I already talked a little bit about the terrible symptoms of having aspiration with chemodenervation because you end up with a flaccid obstructing segment of the cricopharyngeal muscle causing obstruction. But really I think the issue with Botox is if you end up with some of that Botox leaching out and getting into inferior constrictor muscles, and I think this is the most common problem. And so these patients are super miserable. They'll come back right away. They may be coughing with all their foods saying they can't it. And this is like significantly problematic. As you might imagine.

Dr. Greg Dion:

So what happens is, and if you want to visualize this, we talked about the physiology of swallow, your pharynx contracts and your pharyngoesophageal segment open. So in this case, we're supposed to weaken the cricopharyngeal muscle, but if you incidentally weakened the pharynx and not the CP muscle, now you have a weak pharynx against a still increasingly tonic cricopharyngeal muscle, which

can make the swallowing much worse. So, that's really an important thing to think about and probably why the denervation is a little bit less popular now, partly also because we know if we get you better with dilation, you can get people better. Now as far as dilations, well largely these are safe procedures, things that can go wrong and it happens. You can actually take a bougie and get a bougie to actually end up in a ... [inaudible 00:38:03] through a small tear in the serosa and you can get a perforation from a bougie.

Dr. Greg Dion:

You can in theory, get a perforation from a balloon. And then, those are both very uncommon, but important to think about. I think the risk is that you need to do enough, meaning that when you do a balloon dilation, you should see some small mucosal tears. You should see some incidents that you've done stretching. If you seen nothing, you probably maybe didn't stretch enough. And so it's a fine balance, but largely the issue with those conservative procedures is they're not permanent. And so that patient showing back up in your clinic saying like, "Hey doc, thanks for helping me. But I'm back here with the same issue. What do I do now?"

Dr. John Marinelli:

Yeah. And when we talk about complications of CP myotomy, I think one of the most critical ones to consider as is obviously mediastinitis, would you mind touching on that?

Dr. Greg Dion:

Yeah, of course. That's the feared complication, as you're doing [inaudible 00:38:59]. It's one of the first things you learn as an intern on the service is, "Hey, you got to make sure this patient had a myotomy. We need to make sure they aren't at risk for mediastinitis. And the problem is true mediastinitis, meaning you're looking at mediastinal inflammation with leak of all of the pharyngeal esophageal secretions into the true mediastinum is a devastating complication can have upwards of 50% survival. However anatomically we don't always end up there. So we talked about the posterior [inaudible 00:39:32] dissection, as you're making your dissection endoscopically being the buccopharyngeal fascia. So if you've just pierced the buccopharyngeal fascia a little bit, you might only end up in the retropharyngeal space.

Dr. Greg Dion:

So for all intends and purposes, we've talked about retropharyngeal abscess as I'm sure elsewhere. And so you're looking at that and that inflammatory process. If you happen to get through and you've been overly aggressive and you've like tore right through the BC fascia, and now you're through the ALAR fascia and dangerous space. Now you've got a direct communication down there into the mediastinum and now that's going to be very problematic. And so this becomes problematic for a number of reasons. One, it's a hard area to get to, you're talking ...trying to get a drain in there, have a neck incision versus getting a VATs with the cardiothoracic surgeons involved to try to drain that area that's causing inflammation. So it can be a very challenging area to reach. And it's one of those things you have to watch for.

Dr. Greg Dion:

So careful, good surgical technique is a really an important way of minimizing that risk things to think about is by the time the patient's febrile, you're really far down that pathway. One of the first things you're going to think about is saying, "Hey the patient's going to call, this is the classic story." A patient

complaints, the nurse calls whomever at night and says, "Oh yeah, the heart rate's up." And so immediately what's everyone's response? "Oh, that patient's uncomfortable, that patient's in pain." And so they're like, "Well, let's give them, pain meds." But in this case, it's really important to stop for a second and just think about what you're doing and say, "Wait a minute, this patient had a myotomy, do they have other associated symptoms of potential mediastinitis? So aside from the heart rate being tachycardic, are they complaining of chest pain right in the sternum?

Dr. Greg Dion:

So we think about some sternal chest pain and then, understanding what's going on is key, because it's not something we want to rule out. That's where, do we feel crepitus in the neck Is there air in the neck? Is there something that needs to undergo immediate computed tomography scan? Or is this something that we can watch? And it's really important to think about that. One of the things that might happen is, you might be really proud that, "Hey man, that surgery went awesome ... smooth. That tight UES is gone. It's sewed back together, we're all happy." And then for whatever reason, the patient goes into a coughing fit in their room and the post-op area and has a tear through there. And then that's hugely problematic.

Dr. Greg Dion:

Now, interestingly, depending upon how you did it, that might be less or more, or just as much of a problem in the transcervical approach, because in the transcervical approach, some people will leave a drain, others won't. So that's a possibility of draining any of that leakage out. And so the issue is opposite in the transcervical approach because you're coming from the outside in. The question is, did you actually penetrate the mucosal lining? And so that's really the reason we keep these people in house to make sure we don't miss that just because it can be so devastating. With that said, if you take some time and read the nuanced literature, you'll come to see that the reality is most patients whom have a leak get found soon enough that they don't actually develop true mediastinitis. So that's an attestment to what we could say is good doctoring.

Dr. John Marinelli:

Yeah. And the last thing I wanted to touch about in regards to complications or just differences between open and endoscopic CP myotomes, what are the different complications you have to think about between those two approaches?

Dr. Greg Dion:

Yeah. So, great question. When we look at that first and foremost is a perforation, right from the way I just over-viewed that part just a little bit. I mean, if you're doing transcervical, you're talking about perforation from ... perforating through all the muscle and then the mucosa versus the other way where you've perforated through the mucosa, which you knew you were going to do it through the muscle, which you were planning to do, and then the BC fascia into the neck. Either way you worry about things inside the neck. I think that the reality is when we ... where you look back and we take our time to read the best available data, they're telling us that there's really no increased difference with mediastinitis one way or another as a wash.

Dr. Greg Dion:

There is a sense that the subcutaneous emphysema like crepitus would be more common with the endoscopic approach, which makes intuitive sense. But I think the real difference though, is that by

doing it endoscopical, you have a very low chance of injuring the recurrent laryngeal nerve, which is a potential complication when you're making that incision and dissecting through it next. So I think that really is the thing that separates the endoscopic from the transcervical approach.

Dr. John Marinelli:

All right. And transitioning to our last portion. What does long-term followup look like for these patients?

Dr. Greg Dion:

Yeah, so long-term followup really depends on the intervention. So if you've had these patients on a proton pump inhibitor for a month and a half or so, you got to see them back to determine, did that help? Was that the issue? That's one of the reasons that sometimes it's good to just get that objective data and say, "Hey, let's just determine if it's reflux related, let's go and do a PH study. And really there's the ... what's the overall cost of medicine, right? Do we spend some money up front to get the diagnosis and then, we only see the patient twice or do we keep working and trying all these things and seeing the patient multiple times, it's like today or tomorrow, when are we going to dive in?

Dr. Greg Dion:

I think that if you look at someone who's had balloon dilations or Botox or both, you're going to see those patients back. So you often plan ahead. So if you do a Botox patient, you probably should bring them back and four months or so, and just say, "Hey, how's it going? Let's see what you're doing?" Plan a repeat dilation if that's going to be what's needed. And so you get those patients for a while. I have a lot of patients that get a balloon dilations for radiation induced fibrosis. And so these people I know are going to go through a series. I'll plan ahead and say, "Hey, we're going to do this every, six weeks for a few times, see where we get." So objectively look at where we started and where we get to. And you can do that with either a patient recorded outcome measure, like the ET 10 score, or maybe even start to look at pharyngeal constriction ratio on a Videofluoroscopic Swallowing Study.

Dr. Greg Dion:

But, you're going to see those patients back to determine, are they going to need a repeat intervention? Depending upon your practice set up, you can have them reach out to advanced practice provider like a PA or a nurse practitioner, or just call you if they will need to be seen, or you can set something up in the books. It really depends on the setup. And then, one of the advantages, both for the patient and your busy clinic is a cricopharyngeal myotomy patients after ensuring that the [inaudible 00:46:17] after ... really don't need much followup, because they're often satisfied. The things you want to make sure, and that you deal with, coach through and help is any potential increase in the reflux sensation or aerophagia that is not uncommon after doing those myotomes. So I generally follow them up at least to make sure they've healed over the course of a few months before I say, "Hey, high five, we've cured this problem."

Dr. John Marinelli:

Awesome. Well, thanks so much, Dr. Dion, I think this has been a really excellent discussion. Was there anything else you wanted to mention before we transitioned to the summary?

Dr. Greg Dion:

No, John, I really enjoyed being on here and I think that I tried my best to stay out of the numerous rabbit holes that I find myself going down, but, it's an area that I'm passionate about. And I think that as an otolaryngologist, regardless of our exact practice setting be it in a private practice or in an academic practice, there's an opportunity for all of us really to help these individuals. And so the beauty of dealing with dysphasia specifically as it relates to cricopharyngeal muscle dysfunction is you get to do a lot of that doctoring and think through the issue and you can tease out what the situation is and really help these people, that aren't getting a lot of treatment from a lot of other sources and have probably seen a lot of people. So I really appreciate you having me on here and have an opportunity to talk about something that I'm passionate about.

Dr. John Marinelli:

Yeah. It was a pleasure having you. It was a lot of fun having you on the podcast. All right. Well in summary of our discussion today, a description of the classic patient with CP muscle dysfunction is a bit elusive due to the wide variety of etiologies, spanning everything from neuromuscular disorders to esophageal reflux, but classically a patient with symptomatic CP bar is similar to the demographic of Zenker's being more common in older age, and they will present with dysphasia potentially with some other symptoms, if severe, even aspiration pneumonia. Diagnosis requires exclusion of other causes of upper esophageal sphincter dysfunction, and typically includes Videofluoroscopic Swallowing Study that shows an obstructing CP bar in asymptomatic patient.

Dr. John Marinelli:

Conservative treatment options include watchful waiting for the asymptomatic patient, swallowing exercises, such as Mendelssohn's Maneuver and treatment of underlying reflux disease. And there are a couple of conservative interventions, including Botox, as well as esophageal balloon dilation. Oftentimes these interventions are helpful to discern which patients might benefit from definitive management with the CP myotomy and the CP myotomy that can be done either transcervical or endoscopically, and the primary postoperative complication you want to be keeping in mind is mediastinitis. And lastly followup is of course dictated by the type of intervention, but patients who undergo uncomplicated CP myotomes are often very satisfied and do not require longterm followup.

Dr. John Marinelli:

Now we will move into the closing portion of the podcast. While I will ask a question, pause for a couple of seconds and allow some time for you to consider the answer and then give the answer. So the first question for today is, which muscles make up the upper esophageal sphincter? The first muscle is the cricopharyngeus. Second is inferior constrictor muscles, and the third is the proximal cervical esophagus. Next question is, what is typically considered the test of choice to evaluate for CP muscle dysfunction? The test of choice is the Videofluoroscopic Swallowing Study, which will often show an obstructing CP bar. What are the primary interventions for CP muscle dysfunction? The primary interventions are Botox injections, esophageal balloon dilations, or definitive management with a CP myotomy, done either through a transcervical or endoscopic approach. Oftentimes endoscopic has done with C02 laser and just recall CO2 laser wavelength is 10,600 nanometers. And the chromophore is water. Last question. What is the primary complication you should be mindful of following a CP myotomy?

Dr. John Marinelli:

The primary complication is of course, mediastinitis, recall the layers of the pharynx include the mucosa pharyngeal constrictor muscles, buccopharyngeal fascia, that leads into the retropharyngeal space. And

you find that ALAR fascia, which then leads in the dangerous space and finally the prevertebral fascia. Mediastinitis occurs with disruption of the ALAR fascia and infection can quickly spread down the mediastinum and the first clinical sign is often tachycardia patients may complain of chest pain.

Dr. John Marinelli:

Well, that will wrap things up for today's podcast. Thanks so much for joining us and we'll see you next time.