Patrick Kiesling:

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Dr. Alyssa Smith:

Hello everyone, welcome to another episode of ENT in a Nutshell. My name is Alyssa Smith and today we're joined by pediatric otolaryngologist Dr. Doug Sidell. Today we'll be discussing laryngeal clefts. Thanks for being here, Dr. Sidell.

Dr. Doug Sidell:

Thanks for having me Alyssa.

Dr. Alyssa Smith:

So we usually start with presentation. And in regards to laryngeal clefts, how does a patient typically present?

Dr. Doug Sidell:

It's a good question because patients with laryngeal clefts, they can present in a wide variety of ways and many of the symptoms that patients who have a laryngeal cleft present ... These symptoms are common things. Coughing, recurrent respiratory infections, things that look a lot like asthma. They can also have frank choking with drinking liquids or with eating, but a lot of times it's not that clear. So if you think of a opening between the airway and the esophagus you think "Oh, well it's very easy. Every time a kid drinks they're going to have something go in their airway and they're going to cough and it will be that specific. Maybe they'll turn blue." Not always. Cyanosis or color change is possible, but more often it may just be a small child or even a older child who's having failure to thrive and recurrent infections.

Dr. Alyssa Smith:

Do patients always present with symptoms?

Dr. Doug Sidell:

Your question, do patients always present with symptoms? Is a really good one. We have a lot of patients who have symptoms and we don't know they have a cleft, and we ultimately end up finding that. There are sometimes patients who are completely asymptomatic are either coming to the operating room for another reason or we stumble upon the cleft. So it can be quite surprising that patients with clefts small and large can be entirely asymptomatic.

Dr. Alyssa Smith:

So with that in mind, how common is it? And do we even know how common it is?

Dr. Doug Sidell:

In reality, the answer to your second question, I think the answer is no. We like to say 1 to 10 in 20,000 births, which would be something that's ... Not too common. It wouldn't be something that even a pediatric otolaryngologist sees every week. But we see this all the time. So is this likely underdiagnosed? It probably is. The more we start to look for these subtle clefts, the small clefts at the top of the larynx,

or the more we change our definition of what a cleft actually is, these numbers can change. The incidence varies in the literature and it can be as high as 7.8% in some of the studies of kids undergoing laryngoscopy and bronchoscopy for aspiration. Which, if you think about that number, that number's quite high.

Dr. Alyssa Smith:

I think to better understand all of the anatomy and physiology behind these, can we talk a little bit about pathogenesis and what happened during development that actually causes a cleft to form?

Dr. Doug Sidell:

Yeah. With any congenital anomaly, we look at who's having these and we look back at embryologic development. Because in things that are congenital, meaning you're born with it, it's something that goes on before kids are born. So looking at the pathogenesis is important. The first time we actually recognized that clefts occur was ... I want to say over 175 to 200 years ago. And at that point, we didn't really have an understanding of why these things were present in some kids more than others. What happens during the development that causes the anomaly of laryngeal cleft to form has to do with the way that our esophagus and trachea are one big pipe, and then they actually separate during embryogenesis. So if you can imagine your trachea and esophagus having to form a partition between the two and then trying to decide what would happen if part or all of that partition didn't form, you could easily see how these kids could have certain types of laryngeal clefts.

So if you look all the way at the top of the larynx, the cricoid ring, which is the only complete ring of cartilage in the airway, at least it's the only one that's supposed to be there, that ring is totally formed by about the seventh week. So these high clefts, these type one and two clefts that we see, those are usually things that are happening by the seventh week. The clefts that go further down have to do with incomplete development of that tracheoesophageal septum, which can end up with ... That doesn't form. In its entirety you can have a cleft that goes all the way through the larynx into the trachea and down a bronchus, which can be quite devastating.

Dr. Alyssa Smith:

When these patients present to our clinic or we're seeing them on the inpatient setting, what are some of the important questions that we should be asking about as far as their history?

Dr. Doug Sidell:

First of all, I want to say that the more we subspecialize in medicine and surgery, the more we start to understand that patients with complex aerodigestive disorders, which this embodies that, it's an airway and esophageal problem. A lot of these evaluations are best performed by a multidisciplinary team or an aerodigestive center. At Stanford we have an aerodigestive center, so we have our otolaryngologist, speech pathologist, pulmonologist and GI docs who are frequently involved in these patients. And we want to know everything about the baby or the child basically from the time they were born.

And sometimes we have some information from before they're born, so we want to know their birth history, we want to know how have they been growing and developing. If they have symptoms, when did the symptoms develop and what are we doing to try to mitigate those symptoms? Is this just using pulmonary inhalers, are we changing the diet? And with that in mind, what's the child currently eating? Is this somebody who can drink all liquids without problems, or is this somebody who parents have to thick up the liquids to keep their lungs safe? And are they showing up in the emergency room or

in the outpatient clinic or, worse, in the ICU because they're having these terrible recurrent respiratory infections? We want to know all of these things.

And if you really want to simplify it in your mind, think about the anatomy, think about the pathophysiology and the embryology we just discussed, and think what could happen if we're chronically soiling the lungs or if we have a defect between the trachea and the esophagus. The idea is you may have symptoms that present like asthma, chronic wheezing or chronic infectious-like symptoms. But why are they there? And why are they there in that child and not in another child? And that's where the anatomy comes in.

Dr. Alyssa Smith:

So also with that in mind, what are some physical exam findings that we should be looking for?

Dr. Doug Sidell:

Like we mentioned in the beginning, some of these patients have almost no symptoms and we stumble upon the cleft inadvertently. I think that's less common, especially with the larger clefts. The physical exam findings we're looking at predominantly have to do with pulmonary symptoms and pulmonary findings such as abnormality on auscultation? Are these children aspirating? In our clinics we do swallowing evaluations and we actually observe feeds and do clinical evaluations. And some children will actually have stridors, so they could have noisy breathing. And depending on where that cleft is and what's causing the stridor. That can be inspiratory, biphasic or expiratory stridor.

Dr. Alyssa Smith:

And then I'm going to go ahead and assume that, for most of these patients at least, we'll be performing flexible laryngoscopy in the outpatient setting. And when we're doing that, are you able to visualize the cleft, or are there other things that you're looking for?

Dr. Doug Sidell:

Yeah. Again, all that wheezes isn't asthma, and we do a lot of looking in surgery. Part of what we do that separates the pediatric otolaryngologist or the otolaryngologists that matter, from the primary care provider or pediatrician, is the fact that we actually do these procedures in our clinic and we can look at the larynx. When it comes to laryngeal clefts, if you have a significant cleft, what I want you to picture is that esophageal mucosa furling into the airway. And there's something that has been described in the past, and I believe off the top of my head that it was Inglis who described this, but there's something called the [ram sign 00:08:02] where you kind of have a furling, and picture a ram's horns, a furling of the esophageal mucosa between the arytenoids. But it's subtle, you almost have to wonder what's that extra tissue there for? Is it just swelling, is it something else?

Many clefts, especially the high clefts like a type one laryngeal cleft, it's not the most obvious thing on flexible laryngoscopy. And if somebody were to ask you "Hey, is flexible laryngoscopy the tool to diagnose a laryngeal cleft?" It is not. You can miss a lot of them. You may be suspicious for a cleft, there may be findings that suggest a cleft, but it's not the gold standard for diagnosing it. What we do occasionally in the clinic is while we're scoping kids we feed them, and that's a FEES once upon a time that was fiber optic endoscopic evaluation of swallowing, but as time has gone on and fiber optics are less common, we I think are changing that now to flexible endoscopic evaluation of swallowing. And when we're feeding the children, sometimes you can see a very coordinated swallow, there's no evidence of discoordination, the timing of the swallow is right on time, but you are witnessing

aspiration. Whether it's post-prandial aspiration or whether you're seeing actual penetration and aspiration through that posterior interarytenoid space.

But we have many patients who undergo FEES, or this endoscope swallowing evaluation, who have a cleft and they don't have obvious findings on that exam. So I think FEES and laryngoscopy are helpful, but if they're not overtly positive it doesn't mean that a cleft is not there. So sometimes we use other imaging modalities like modified barium swallow study or video fluoroscopic study is another word for that, which is, again, your MBSS or your VFSS, to further evaluate the swallow. Now remember, the difference between that and a FEES is that the VFSS can actually show you what's going on during the pharyngeal contraction phase of swallows. So when the pharynx completely whites out our scope because it contracts around it, you can see that on an x-ray swallow test. These two tests are complimentary, you think of it like a Venn diagram, they both have a little bit of overlap but they both have their own things that they can show you.

A laryngeal cleft, people have described a very specific posterior, horizontal penetration pattern. But again, like the FEES, if this is negative or if it's non-specific, it may not rule out a cleft necessarily, or it may not necessarily rule one in. But you can be more suspicious of a cleft with a subset of findings that you can see on the video fluoroscopic swallow study. So I think that that is helpful. Another thing that a video fluoroscopic swallow study really brings us is ... And a FEES, for that matter, is if the child has symptoms of aspiration and you're thinking "Maybe this kid has a cleft." And you look in there and you're seeing extremely discoordinated swallow, the timing is off, they're having a lot of liquid get down to the hypopharynx or around the larynx before their body's actually triggering that swallow, that's suggestive of somebody who's going to have swallow problems probably no matter what, even if they have a cleft. If you have a cleft and you have that going on and you fix it, you may not be as likely to fix the whole problem because there's a coordination issue as well.

Dr. Alyssa Smith:

And then I know for a lot of kids that have congenital anomalies, there are some associated syndromes that we should be thinking about. How about for laryngeal cleft, are there any associated syndromes that we should be considering?

Dr. Doug Sidell:

Yeah, absolutely. It wouldn't be ENT without a syndrome tied to it. Pallister-Hall and Opitz Frias are probably the two biggest ones, and those are the ones that we see classically on our board exams. But in practice, I think that based on the rarity of some syndromes, we will see in our practice things like VATER or VACTERL association, CHARGE association as well, so with those I think it's very important to be looking for or at least thinking of a cleft. Also in patients with other associated GI or genitourinary, cardiac craniofacial abnormalities, we need to be thinking of laryngeal cleft as well as other midline defects. Cleft palette, tracheoesophageal fistula, et cetera.

Dr. Alyssa Smith:

Do you refer these patients to genetics when you see them?

Dr. Doug Sidell:

Not always. I think if we have an isolated problem, no other signs of syndromic abnormalities, we don't necessarily need to send them to genetics. That being said, a lot of these patients end up with genetics anyways, and that's usually by virtue of some other things we're finding on some physical examination.

Dr. Alyssa Smith:

And then moving further down our workup and evaluation in the operating room, can you walk us through how you do your evaluation in the OR?

Dr. Doug Sidell:

Absolutely. Again, in the past in this lecture series I mentioned that a flexible laryngoscopy is not the gold standard. So that begs, what is the gold standard? The gold standard evaluation to diagnose a laryngeal cleft is a rigid endoscopy with palpation. Now you don't need to use a ventilating bronchoscope by any means, but what you should do is you should get a laryngoscope into the airway and then you should feel that interarytenoid space. Whether you suspend the laryngoscope and feel, whether you place a cord spreader or not, I think there's a lot of controversy about that. Sometimes if you put a laryngoscope in ... And then you suspend it, you can actually rotate the posterior aspect of the larynx anteriorly.

So it can almost take a very subtle cleft and hide it. Whereas if you're just putting a laryngoscope in and holding it with your hand, you can see and feel with something like a right angle probe, that you do have a space between the arytenoids that extends all the way down to and/or below the level of the vocal folds. So it's very important to look with a rigid instrument and feel. The cord spreader ... It shouldn't be called a cord spreader. A laryngeal spreader or a false vocal cord spreader is something that can splay the vocal folds laterally and allow you to really scrutinize that interarytenoid space. So that's another instrument that's frequently used.

Dr. Alyssa Smith:

What are some important features of the cleft that you are evaluating?

Dr. Doug Sidell:

I want to know how obvious the cleft is just on direct visualization. I want to know how deep the cleft goes. So one of the things that we're seeing a lot of now is practitioners will go in there and they'll see some space between the arytenoids that looks deep. Sometimes a space actually looks deep because they have tall arytenoids, and the classic example is somebody with laryngomalacia who's diagnosed with a laryngeal cleft, well, that's not always a laryngeal cleft. They may just have laryngomalacia tall, floppy arytenoids. So you really have to feel, and then you have to use your right angle probe to really compare in that plane how deep that cleft is. And then you also, like I mentioned before, you're feeling.

So some of these clefts are subtle, and if you feel that interarytenoid space and you actually put that probe there, it will actually sink deeper than you would expect with your naked eye for that probe to sink, and then you feel the cricoid. Is there a notch in the cricoid? Does this cleft actually extend into the cricoid cartilage? And you'd be surprised at how many kids who have a type one laryngeal cleft, you feel that and you actually realize "Nope, this actually extends partway into the cricoid cartilage," That's a type two. And we can go over some of the different types in a little bit.

The other thing that you have to be careful of when you're feeling for these is that you don't really just push hard on that interarytenoid space, because you can make anything look like a cleft. The interarytenoid musculature and the interarytenoid soft tissue is soft. So you can push that in and actually put that probe right down to the level of the vocal folds. That does not mean you have a cleft.

Dr. Alyssa Smith:

And then can you just speak to the importance of the comprehensive airway evaluation when you're in the OR looking for something like this?

Dr. Doug Sidell:

Sure. One of the more embarrassing things you could do is go in there, find a cleft, get out of there and then realize you missed one or even two tracheoesophageal fistulas. What about anomalies and bronchial branching patterns? It's worthwhile in a multidisciplinary evaluation to actually do a pulmonary bronchoscopy as well. Paul Boesch and Karthik Balakrishnan wrote a paper discussing the utility of flexible bronchoscopy. They actually in their hands showed that it's a capable means to identify some of these clefts in experienced hands. Now, it's not the gold standard, but it is something that is frequently done. Something meaning a pulmonary bronchoscopy. But something that's also done during the evaluation of a patient, say, with aspiration symptoms. So doing a thorough pulmonary and tracheal airway evaluation is extremely important.

Dr. Alyssa Smith:

And so far we've mentioned the different types of clefts that can be present, but can we just go over the grading system that we use?

Dr. Doug Sidell:

Absolutely. The classic grading system, and I think that anybody today would probably go back on that Benjamin Inglis classification system, because it's the most commonly used classification system. The type one cleft really just goes down to and maybe just below the level of the vocal folds. The key with this and what distinguishes a one from a two is it does not go into the cricoid cartilage. There's not a notch in the cricoid cartilage, it doesn't extend, necessarily, too far below those vocal folds, so that's why feeling is so important. So type one pretty much to the level of the vocal folds, type two, into the cricoid cartilage. Now type three, if that cleft goes through the cricoid cartilage, now you're in trachea. So if you go through the cricoid cartilage, that is a type three. You're in the trachea but you're still above the sternum, above that breastbone. So you're in the cervical trachea.

So what makes a difference between a type three and a type four? All right. Type four, now you're in the thoracic trachea. You're between that area where the thoracic inlet starts in the carina. Some of these, like I mentioned before, will extend all the way into a bronchus, and those can be a bear to fix and most of those kids have symptoms right after birth. There's also this difference in opinion out there about what is a type one laryngeal cleft? Because remember, I said it's above the cricoid but it's down to the level of the vocal folds. Where does that start, what's the upper boundary of that? And everybody has a different opinion, so we've started using this phrase deep interarytenoid notch. I think if you're going to be out there describing a cleft, just describe the interarytenoid space. If it's above the cricoid cartilage, just say the interarytenoid notch goes down to the level of the vocal fold or to the level of the ventricle or to the level of the false vocal fold.

If I have a interarytenoid space that goes to the level of the false vocal fold, that is not a cleft in my eyes. It's not something that I'm going to fix. But if I have a patient who's aspirating and a space that goes down to the level of the vocal folds, sure, I can call it a type one laryngeal cleft or I can just describe why I'm in the operating room doing that procedure and make a decision with the family as to whether or not to fix that. There are also these occult clefts, which are clefts that may have a mucosal covering so they're not as obvious. But when you feel them, you'll actually feel defect in the cricoid ring. It's not something that's talked about as much, but if you're out there in practice it's really something to remember. Occasionally kids can have some mucosal defects in their laryngeal superstructure.

Dr. Alyssa Smith:

Now that we have our diagnosis and we're looking at management or treatment, what exactly are the goals of treatment for these patients?

Dr. Doug Sidell:

Right. They're coming in with symptoms many times, and remember we talked about they're having pulmonary complications and may have difficulty swallowing? You want to prevent that. And in some cases, which is more uncommon, you have stridor. So that's airway obstruction, that's air passing rapidly through a narrow space, usually in the larynx or trachea. So if you want to reduce stridor you're reducing airway obstruction. Again, less common but it can happen.

Dr. Alyssa Smith:

And what is the treatment approach for these patients, and can any be managed conservatively?

Dr. Doug Sidell:

Absolutely. I think the reality is type one clefts, we really are relying on the function of the patient. There's been a couple papers out there, but one of the patients in Boston described about a 20 to 30% incidence of type one laryngeal clefts that are diagnosed that will be okay with conservative management alone. I think that if you have a laryngeal cleft that's a type two through four, we're frequently fixing those because of the known risks of having the cleft over the long term, as well as a lot of the other issues that justify repair in these patients, whether they're syndromic or they have other medical problems that really require us to keep the kid as spruced up as possible. But there are patients who can just get by by thickening the liquids, and we can also control reflux to make sure that we're not having additional inflammation or reflux and associated aspiration.

Not every kid can be thickened, and every hospital has different rules as to how old do you need to be before you start using some of these thickening agents and which thickening agent to use. And there are some kids who, from a medical risk standpoint, sure, you can thicken it, but they're not going to drink it. So when you see your patients, remember, you're looking at individual kids and you have to work with the resources that you have. So while you may have two children that look very similar on swallow study, have a very similar microlaryngoscopy and bronchoscopy, one patient may tolerate thickener and the other one might not, so you may be a little bit more likely to fix the one who doesn't tolerate the thickener.

Dr. Alyssa Smith:

And before we jump into surgical repair, can you talk a little bit about the role of interarytenoid injection?

Dr. Doug Sidell:

Sure. If you go on PubMed and you type in interarytenoid injection, you're going to find a lot of papers that talk about injecting clefts, injecting every interarytenoid space, et cetera. I think everybody has a different opinion as to how well this works. I think that patients who are okay candidates for this if injection is part of your practice are really only patients who fall in that type one cleft category. I don't see a lot of utility in injecting a type two laryngeal cleft. Foremost, the injection is going to give you a little bit of bulk in that interarytenoid space, but it's not going to give you nearly as much closure of that interarytenoid space as, say, a suture technique. In addition, if you inject that area and it doesn't work,

does that mean that closing the cleft isn't going to help? No, it doesn't mean that. Because like I said, you're not doing as significant of a closure with an injection as you are with a surgical closure.

Now, that being said, if you do an injection and the patient actually has a dramatic improvement in the symptoms and as that injection material wears off the symptoms come back, that may justify you doing a surgical repair because what you're shown yourself or what you've supported, at least, is that that notch or cleft is actually contributing to the problem. You get rid of it one way or another, it is going to help. So that's, I think, where injection falls. In my personal practice, if you want to know how often I inject, very little. At the beginning of my practice I injected more often, and it's not a risk free operation, it's a low-risk operation. But it's not a risk-free operation. So I don't inject too often anymore. I usually, if there's a cleft there and it's the problem, I just sew it shut.

Dr. Alyssa Smith:

And what are some materials that are commonly used? Does it differ from kids versus the typical injections that we would use for adult vocal fold paralysis?

Dr. Doug Sidell:

I do a lot of vocal fold augmentation, and I think that a lot of these are very similar substances. Hyaluronic acid is, I think, a good one if you're going to do something, Juvederm is an example. Those will last in the range of months. There are also certain types of gels you can use. One of them is Prolaryn gel, or the carrier gel that was used for calcium hydroxylapatite. The old name was Prolaryn Gel and Prolaryn Plus. Now, the carboxymethylcellulose, that lasts a matter of weeks. It's almost like a gelatinized water. It is probably the injection with the lowest side effects, but probably the lowest effect. There are things like gel foam, which are purified pork skin gelatin. They use gel foam for things like causing coagulation. Those will tend to last six to ten weeks, I'd say.

People were using that for vocal fold injection as far back as the 70's. Cymetra Micronized Alloderm, that again will be in your range of months, if not longer. I think some opinions are mixed about that. And then some people who have written about this have talked about calcium hydroxylapatite, which is ... Picture a gelatinized cement being injected back there. If you misinject this it's not going away anytime soon and you could actually cause more problems than not. So if you're going to inject something I would pick something like hyaluronic acid, something that you're comfortable with, it's pliable, you can remove it if you need to but it's not going to last forever.

Dr. Alyssa Smith:

Let's talk about the repair. Can you discuss the surgical approaches that you use?

Dr. Doug Sidell:

Absolutely. The first open approach to ... Remember, I said this was first identified 200 years ago. But the first time we actually said "Let's cut somebody open and fix this." Was in 1950's, I think '55, and that was Petterson. Things have changed dramatically at the time. I think he was closing a type one or a high type two laryngeal cleft. There's a bunch of different approaches for the various types of clefts. The big dividing line is do you do it open or do you do it endoscopic? So type one clefts, again, we frequently rely on function as to whether or not we're even going to close them. But if we do close them, I would have a really hard time justifying an open approach for that. That is an endoscopically repaired surgery. The type two through type four, a lot of times these patients are going to the operating room no matter what. The question is whether or not you're going to do it endoscopic or open.

Type two and many type threes, I think I used to say some, but many type threes actually can be done and done well endoscopically. And then ... The long type threes and the type fours is best treated with an open technique. So if you're comparing these endoscopic versus open approaches, the endoscopic approach, there are a lot of different opinions as to how you should do this and what instruments you should use. All patients will require a suspension laryngoplasty, so you insert your laryngoscope of choice, you suspend the larynx, you bring in an operating microscope, and then you have to decide am I a cold steel guy or am I a laser guy? Personally, I use cold instruments because I think it teaches the residents a lot about laryngeal instrumentation, but I'm fine with a laser as well. So use what's good in your hands.

The idea is what you really want to do is denude all of that mucosa throughout the cleft. And the big place people like to leave mucosa behind and cause problems is the apex. So put a vocal fold spreader in there, a laryngeal spreader rather, spread that open. I do a little bit of an injection with a little oral tracheal injector using some Lidocaine with Epi. Let that sit for a sec, then I denude the mucosa with cold steel and then I do, personally, a mass closure technique. So I use a PDS suture, usually a 4.0. I do it with an RB1 needle, you can use a 5.0 with an RB2 if it suits you, you can use Vicryl, it lays flat, I just personally don't use it. And I take the going from posterior to anterior, left to right. I will throw sutures starting at the apex and working back. You really need to make sure that apex is completely closed.

Other people will actually do this in layers. They'll sew that anterior mucosal layer and posterior mucosal layer separately. I think that takes more time and the studies that we have done shown there's not any difference in the outcomes and both dehiscences and/or swallow up comes for these patients when you think about it. Open approach, there are a bunch of different ways to do this. In the old days we talked about these lateral pharyngotomy approaches, they're not really used anymore. The steps of the repair for these really depend on where the cleft is, so you can do an anterior approach, picture making an incision in the anterior neck, going down onto the trachea and the larynx, making a split through the cricoid and at a bare minimum the inferior aspect of the laryngeal cartilage. A lot of people used to open the entire larynx and do a complete laryngofissure and you may need to do that for your reach. Remember, you're sewing up this entire arytenoid space as well as the cleft itself.

When we do this we take the esophageal mucosa, and a lot of times that's redundant mucosa. Remember I told you that you can have stridor with these clefts? What's happening when you have stridor is a lot of that esophageal mucosa is flopping into the airway. A classic mistake, just as an anecdote, that I made when I was a fellow, is I had a patient with a type three cleft. Miraculously he wasn't aspirating. But we identified the cleft and what was the first thing I did? I put in a nasogastric tube. What did that nasogastric tube do for this little baby who was breathing on room air? The nasogastric tube pushed all the esophageal mucosa into the airway, it obstructed the airway, and then I had to intubate the patient. So remember that that mucosa's there and it's redundant.

When you're doing these repairs, one of the first steps you do is you decide how much extra esophageal mucosa do I have in the open approach, and how much can I trim? So you're trimming this in layers, you're separating that tracheal component from the esophageal component, and you're closing the esophageal component and then we use an interposition graft, which I can talk about a little bit more in a sec, and then we close the tracheal segment. Some people do this with running sutures, some people do this with an interrupted suture and buried knot. What I try and do is I try and keep the suture and knots out of that space between the trachea and the esophagus. I have no problem if I have a couple knots running up the [alumanumide 00:30:19] trachea and these kids. It has never once caused a problem for me and my mentors, such as Mike Rudder, have done it that way for a long time and it causes no issues.

There are a subset of kids who have these long type four clefts, and I'd be happy to talk about those as well. The type four clefts are, again, clefts that go into the thoracic trachea. There's a subset of these kids who you can actually reach that apex through the neck, and that requires pulling the trachea up a little bit through that anterior approach. Many of these kids, potentially, and especially if it goes down the bronchus, the cleft goes down the bronchus, you will need a sternotomy. So you're going to open up ... The chest via sternotomy approach and approach the trachea that way. Now whether you make a linear cut down that anterior aspect of the trachea and approach it straight through the trachea, sewing the esophagus and trachea and causing that separation that the patient needs.

Or you do a different type of approach, which is actually one that I do for type four clefts, and again, this is something that I learned from my mentor Mike Rudder, where you actually do a laryngotracheal separation of sorts where you ut the trachea off of the larynx at the level just inferior to the cricoid, and now you're looking down the lumen both of the airway, but you're also seeing into the esophagus. Then you make a separation between that mucosa of the esophagus and the trachea, and then you sew from the apex, that distalmost point of the cleft, all the way up proximally to the interarytenoid space. You do it for the esophagus first, you lay down that interposition graft and then you close the trachea. Then you [inaudible 00:32:06] the trachea to the larynx. So that is an excellent technique.

The one last thing I'll talk to you about for these long clefts where you're doing an open approach, let alone a sternotomy, I actually put those type four clefts on bypass pump or ECMO. And the reason why is because this gives you the luxury of having the time and the space without a breathing tube in there to close your esophagus and your airway in a very watertight, as they say, fashion. You can do it with a breathing tube and ventilate the entire time, but if you have a long cleft what you need to do is actually put your breathing tube in the airway, and you may have to mainstem one of the bronchi to do it. And then you can suture the tube in place so when you lift up that trachea and you're working on the esophagus, the endotracheal tube doesn't move around, it actually stays in position. It can be quite a bear and quite a nuisance to have that endotracheal tube in the way, so I have done these on bypass and it's actually a quite short bypass run if you need to do it.

Dr. Alyssa Smith:

You mentioned these interposition grafts. When do you use them and what are you using for your grafting material?

Dr. Doug Sidell:

Sure. Again, the interposition are for the open approaches. I think that sternoperiosteum is a very good choice because if you're entering the neck it's right there. If you're entering the chest you just take some before you make your sternotomy, that's the best way to do it. Tibial periosteum is something that may also be used. It's a separate incision on the leg. In the old literature and still occasionally used, you can use plural flaps or sternocleidomastoid. So those are all options.

Dr. Alyssa Smith:

And then how about any surgical techniques to decrease the risk of postoperative fistula formation?

Dr. Doug Sidell:

I think really making sure that you make your cuts on your esophageal mucosa such that ... Like I said, you've got a lot of extra esophageal mucosa. You really want to denude that area so that you have a very raw surface that you're sewing together. For these open approaches you want to make sure that

you're really scrutinizing your suture line as you're throwing these sutures down, you're not leaving gaps, you're not putting things on tension where tension shouldn't be, and you're not overdoing it with the amount of tension you're putting on it. You don't want it to constrict the mucosa or devascularize the mucosa by excessive tension on each knot if you're placing a interrupted or even a running suture. For the endoscopic approaches, again, denude everything and focus on that apex.

When we see these things break down, yeah, you may lose a stitch up high, but that's not a big deal. You throw another stitch at some point in the future. But if you have that apex that opens up, what you end up with is a form of a hole almost like a TEF. It's harder to fix and you may have to open everything up to completely close it if it's causing a problem. So I think that to decrease the risk of fistula formation and fistula development is know your technique, use a technique that works well for you, and with the open approaches just really scrutinize what you're doing while you're doing it. It's really easy to skip layer or lose track. And again, I think that interposition graft is a fantastic extra layer to reduce fistula formation.

Dr. Alyssa Smith:

And then any other surgical pearls or words of wisdom for learners?

Dr. Doug Sidell:

Yeah. I think that the laser versus cold and layered versus mass closure, you do which is best in your hands, and that may depend on your training. Don't be afraid to change if you need to. Also, like I mentioned, for long clefts consider cardiac bypass. And then I think that the other thing that people tend to forget to do with the endoscopic approach that I've at least witnessed a couple times myself is they will go in and they'll do an endoscopic cleft repair, but what you're doing when you do that is you're bringing the arytenoids closer together. And if you have even the slightest amount of laryngomalacia, what happens is those short aryepiglottic folds, pull the epiglottis down and covers up the airway. So by all means, cut the aryepiglottic folds. Do your little aryepiglotticplasty ahead of time. It'll make not only your closure easier, but it will reduce your stridor post-op.

Dr. Alyssa Smith:

So speaking of post-op, how are these patients managed? Are they discharged same day, do you keep them overnight?

Dr. Doug Sidell:

Personally I don't discharge these patients at all on the same day. Could they go home? The number of complications we've had in the post-op arena with patients with these short laryngeal clefts, type one cleft closures, is very, very uncommon. It's very low. Patients with longer clefts, these type four clefts, a lot of these kids are going into the ICU maybe with a breathing tube in for a little while. The short clefts I tend to put in the ICU just because at my hospital this isn't an excellent place for airway observation. If you have a complex airway unit at your institution, that's a great place for it, or if you just have a very secure and safe floor for patients who don't have any other significant comorbidities and have relatively healthy lungs, I think they could go to the floor without an issue. There are a couple papers that have been written about this. I think there was a Boston paper that talked about a very specific algorithm as to how they determine what patients go to the ICU versus floor, and it really depended on their pre-op evaluation and their comorbidities.

Dr. Alyssa Smith:

And then how about their typical post-op diet?

Dr. Doug Sidell:

If they're not intubated and they're ... Again, with these type one laryngeal clefts, I have them spontaneously ventilating, and type two and three. I have them spontaneously ventilating the entire time. So we tend to go to sleep without a breathing tube and we wake up without a breathing tube. These kids can eat post-op. I think that the type one and two clefts at a minimum, this is a standard for us, I allow them to eat the same consistency diet as they had, same consistency liquids as they had pre-op. If they are on a regular diet I will have them eat a soft diet for a week or two. I'm not as strict about that as I used to be.

If patients are drinking and let's say they were on nectar thick liquids pre-op, we do a laryngeal cleft repair, and now they're coughing a little bit with the nectar thicks, I'm not against thickening it up just a little bit, because they can be sore after surgery. We may deinnervate them a little bit, and they do have some changes to get used to. So I may thicken up the liquids a little bit. I do give them reflux medication after surgery and I avoid steroids if possible.

Dr. Alyssa Smith:

What are some post-operative complications that residents should be aware of when we're managing these patients?

Dr. Doug Sidell:

This is one of, I think, the most important things that you asked me. Because every complication that you think about that may occur to the child is something you need to talk to the family about beforehand. So after surgery you may be going in for aspiration, they may persistently aspirate. I told you about the laryngomalacia and I told you about how we're bringing the arytenoids even closer together or that interarytenoid space. You may even have a little bit of stridor or airway obstruction afterwards. I think that that's something that if it happens it's brief and I've never had to take down a cleft repair, that's for sure. You could have cleft breakdown, and that may happen in two weeks, that may happen in two years.

So a reevaluation in the operating room may be needed. You could have leaks at your anastomotic site, whether you're talking about an esophageal closure or a tracheal anastomosis. You could have tracheal issues if you do a laryngofissure or tracheal transsection, anything that could go along with that including dysphonia and/or tracheal stenosis. You could have strictures [inaudible 00:39:58] the esophagus and you can injure the recurrent laryngeal nerve, depending on what your technique is and how you're doing the operation. And again, these are all things that we need to counsel our families on before repair.

Dr. Alyssa Smith:

Speaking of counseling parents, how do you counsel them on the chance of a successful surgery, and how do we even define surgical success?

Dr. Doug Sidell:

I go through the risks of the outcomes and I talk to them about their child's pre-op swallow evaluation. If they have baseline gross discoordination, the cleft repair may be less likely to be successful. And I talk to them about that. If they have a very focal problem on their swallow evaluation and they have a very

obvious cleft, we may be more successful. There are other things, comorbidities, neurologic issues that may be occurring alongside the cleft that may make their outcomes different than somebody who doesn't have any comorbidities whatsoever. As far as the longterm success is concerned, I feel that it's not too hard to keep that cleft closed, but how successful are we at actually improving the swallow study?

Alex Osbourne was an attending at Cincinnati Children's Hospital. He looked back at a bunch of laryngeal cleft repairs and their swallow studies, and about 57% of the kids had normal swallow function. And then you can break it down that about 20% had some penetration and aspiration. But the vast majority of the kids got better. [inaudible 00:41:29] at Boston Children's also looked at a bunch of their patients, and they had improvement in swallow and all the patients for all cleft types. I think that I can never give a guarantee like that. I think the majority of my patients, I tell them that "We're looking for improvement. I want to be able to thin your liquids down, I want you to have fewer respiratory complications, I want you to have fewer respiratory infections."

Some kids are on nectar thicks but they're still getting in the hospital and having intermittent issues, and we do a cleft repair and their swallow study may look similar, but they're much, much better. Parents are saying "Look, I'm not using my Pulmicort anymore, we don't even have to use our inhalers, and I don't care what the swallow study said, my child is better." So we're looking for improvement on all fronts, and obviously we want a patient who's aspirating to be not aspirating on the swallow study and everything to be perfect. But it's not always that successful.

Dr. Alyssa Smith:

So when we're getting repeat swallow studies after surgery, do you get them for all patients? And what does that follow-up look like?

Dr. Doug Sidell:

I tend to see my patients a couple weeks after surgery just to see how they're doing, they all have my phone number and cellphone, so if issues come up beforehand I'll see them. But I make sure that we're north having any obvious issues and then I tell them I'm going to order another swallow study, for most kids, at about six weeks. And this is important, I think, for those silent aspirators, the kids who aspirate and they don't cough when they have aspiration. And we can see that on swallow studies.

So if we do a cleft repair I'll do the swallow study at about six weeks post-op, and for the couple days before the swallow study, for better or worse, I do have them gradually thin the liquids a little bit so the first drink they're having at the time of the swallow study isn't the first time they've had thin liquids in a year. That's my approach. Now, if the kid is one of those kids who classically coughed every single time they had an aspiration event, and we do a cleft repair and now they're not coughing anymore, I will occasionally follow those patients on an outpatient basis without repeating the swallow study.

Dr. Alyssa Smith:

So thinking about the natural history of this anomaly, what if we didn't repair it? What can we expect to happen in that child?

Dr. Doug Sidell:

I think many kids who have unrepaired clefts, and if we're assuming that the cleft is associated with their aspiration, you can have long term sequela that's associated with your lungs. The bronchiectasis and other pulmonary complications may occur even in the absence of recurrent pneumonias. You may just

be subtly aspirating for long periods of time and ultimately suffer the consequences of that. Patients with larger clefts tend to typically follow a more classic pattern of obvious aspiration, but not always. So if you have a laryngeal cleft and you have some swallow dysfunction and you're not able to mitigate some of the issues that you have, whether that be recurrent infections, chronic inhaler use, chronic inflammation, bronchiectasis, if you're not able to mitigate that with thickened liquids then a cleft repair should be done.

Dr. Alyssa Smith:

Alright. In summary, patients with laryngeal clefts can have some common presenting symptoms such as coughing, choking, dysphasia and occasionally cyanosis as well as failure to thrive in recurrent respiratory infections. Laryngeal clefts form due to failure of the fusion of the posterior cricoid lamina as well as failure of the formation of the tracheoesophageal septum. Workup includes a comprehensive history and physical exam as well as laryngoscopy and evaluation of swallowing function with FEES or video swallow study. Rigid endoscopy with palpation under general anesthesia is the gold standard for diagnosis. Management approach should be individualized and based on symptoms and type of cleft. Treatment includes conservative management with thickening of feeds, as well as a variety of surgical options ranging from endoscopic to open approaches. Dr. Sidell, thanks again for joining us. Is there anything else you'd like to add?

Dr. Doug Sidell:

I really appreciate the opportunity, and I think that the last thing I'd like to say is a couple points. An upfront conversation with the parents is key, especially with these type one laryngeal clefts. Everybody manages them different. The question is, how much is that cleft playing a role in the patient's problem? It's tempting to fix everything, but look at the overall child. What else do they have going on, what's the resource burden, what are you actually trying to accomplish and what are the parents' expectations? Because if what you're trying to do and what the parents want to happen is different, then you've got yourself a problem there that needs to be solved before you fix anything. Again, thanks a lot for the opportunity, I appreciate it.

Dr. Alyssa Smith:

All right, awesome. I'll now move on to the question portion of this podcast. As a reminder, I will ask a question, pause for a few seconds, and then give the answer. The first question is, what is the grading system used for laryngeal clefts? The grading system used for laryngeal clefts is the Benjamin Inglis classification system. This is at least the most commonly used classification system, although others are also present. So in this classification system, a type one laryngeal cleft goes down to the level of the vocal cords. Type two falls below the level of the vocal cords and into the cricoid. Type three goes completely through the cricoid and into the cervical trachea, so still stays above the sternum. And then type four goes into the thoracic trachea.

The second question is, what are some syndromes that are commonly associated with the laryngeal cleft? The two most common syndromes that we think about are the Pallister-Hall and the Opitz Frias syndromes. Pallister-Hall can also be associated with a bifid epiglottis, polydactyly, syndactyly, hypothalamic [hemeratoma 00:47:26] imperforate anus and kidney abnormalities. Opitz Frias is commonly associated with hypertelorism and hypospadius. The two other syndromes or sequences that we want to be thinking about include VACTERL and CHARGE.

The third question is what are some common presenting symptoms for patients with a laryngeal cleft? Some patients may be completely asymptomatic, although when they do present with symptoms

some common symptoms are coughing, choking, dysphasia, recurrent respiratory infections, failure to thrive and sometimes cyanosis. More severe clefts such as a type three or type four laryngeal cleft are more likely to present earlier and with more severe respiratory symptoms. That's all for today's episode, thanks for joining.