

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. Laura Ovidas. Today, we'll be discussing laryngomalacia. Thanks for being here, Dr. Ovidas.

Dr. Laura Ovidas:

Well thanks for having me. It should be fun.

Dr. Alyssa Smith:

First, let's start by defining laryngomalacia.

Dr. Laura Ovidas:

Well laryngomalacia in general is congenital softening of the tissues above the glottis which causes a collapse of the tissues into the area of the glottis causing inspiratory stridor or inspiratory noise. It is the most common cause of noisy breathing in babies.

Dr. Alyssa Smith:

So how does a patient with laryngomalacia typically present?

Dr. Laura Ovidas:

Well we see probably the vast majority of them in the outpatient setting where they've been evaluated by their primary care provider and were noted to have noisy breathing, but occasionally we can also see them in the hospital setting where a child may be hospitalized for other reasons or gets acutely ill and their laryngomalacia is intensified to the point where they're asked to be evaluated while we're in the hospital setting. They usually present shortly after birth. In general, two weeks is kind of the rule of thumb, but whether the severity warrants any further evaluation is kind of what we need to evaluate as both primary care physicians and pediatric ENT physicians.

Dr. Alyssa Smith:

What symptoms do patients typically present with?

Dr. Laura Ovidas:

The main one is the inspiratory stridor. It can be positional. Often laying down is worse or after feeding is worse. Parents will notice it sometimes continuously, sometimes very intermittently. Sometimes one day they'll have it and another day they won't. Often going along with that are things like retractions. If severe, they can also present with failure to thrive. This is usually a result of working so hard to breathe that most of your calories are going to breathing and not growing.

Dr. Alyssa Smith:

You've mentioned this stridor and noisy breathing. What happens in the airway that causes these sounds to occur?

Dr. Laura Ovidas:

Well it's basically a collapse of the tissues in the airway, the area above the glottis otherwise known as the supraglottic structures. These tissues dynamically collapse into the supraglottis and narrow the airway which causes that turbulent flow of air and the noise.

Dr. Alyssa Smith:

So do patients always present with symptoms?

Dr. Laura Ovidas:

That's probably the main way we look for laryngomalacia. It's because of noisy breathing. Now often, patients will present with noisy breathing that really isn't stridor, but because sometimes the primary care physicians have a difficulty discerning what the noise implies or is demonstrating. We'll see things like stertor which is often just nasal congestion. By definition, laryngomalacia causes inspiratory stridor. So when they breathe in, they make the noise.

Dr. Alyssa Smith:

You mentioned that laryngomalacia is the most common cause of noisy breathing in infants, but do we know how common this actually is?

Dr. Laura Ovidas:

Of the kids with noisy breathing in infancy, it's about 60% to 70% of those kids have laryngomalacia and about 90% of them won't require any kind of surgical intervention.

Dr. Alyssa Smith:

So moving on to pathogenesis, what causes the collapse seen in laryngomalacia to occur?

Dr. Laura Ovidas:

Unfortunately, we don't absolutely for sure know. General consensus at this point is it's more of a neuromuscular generated hypotonia probably caused by the sensory stimulation of mechanoreceptors and chemoreceptors of the superior laryngeal nerve at the level of the aryepiglottic fold. There's actually been some histologic studies showing there is an increase in the size of these nerves in children with laryngomalacia. This dysfunction can occur anywhere along the afferent or efferent pathway of the vagally mediated laryngeal adductor reflex, and it causes, in general, decreased laryngeal tone which causes the collapse which leads to the noisy breathing.

These kids also often will have decreased sensation in this area. There's several theories as to that as well. Lot of it is probably associated with GERD and the fact that the irritation from the excess stomach acid being refluxed onto the area causing edema, but there's also thoughts that this increase in nervous generation also leads to some of this decreased sensation in the area.

Dr. Alyssa Smith:

Next, focusing on workup of these patients, when we're called to the bedside to evaluate an infant with stridor, what should be in our differential diagnosis?

Dr. Laura Ovidas:

Well first of all, you need to discuss what type of stridor they have. Certainly like we said, laryngomalacia by definition is an inspiratory noise, not an expiratory noise or a biphasic noise. When you start looking at some of the other possibilities such as subglottic stenosis, vocal fold immobility, cysts, hemangiomas, vascular rings, et cetera, the noise can be described differently. Sometimes it's biphasic. Sometimes it's expiratory. In general, you can talk about things that occur at or around the level of the glottis as being biphasic and things that occur in the subglottis in the trachea are usually expiratory noises.

Dr. Alyssa Smith:

What are some important history questions that we should be asking during our evaluation?

Dr. Laura Ovidas:

Again, you're looking at how the baby's doing. Are they growing? That's a big one. How old were they when the symptoms developed? What's associated with this noise? Are they retracting? In other words, is the area at the suprasternal notch area sucking in every time they breathe? Are they using their abdominal muscles to breathe? Is there any cyanosis? Are they having trouble feeding, coughing, choking with feeding? A lot of those are signs and symptoms of laryngomalacia, and depending how bad it is determines whether or not further intervention would be required.

Dr. Alyssa Smith:

And then when we are performing our physical exam, what should we be looking for?

Dr. Laura Ovidas:

Well the main way to examine a baby for laryngomalacia requires nasopharyngoscopy. You have to actually look at the larynx and it's pretty easy to look at them while ... While you're looking at them, they're often crying and give you a very good view as to what's happening when they're worked up which is a good time to evaluate what the condition of their larynx is. So that's pretty much the gold standard, nasopharyngoscopy.

Dr. Alyssa Smith:

And what are some specific findings that we should be looking for on nasopharyngoscopy?

Dr. Laura Ovidas:

Well it's basically prolapse of that soft tissue. Often there's redundancy of arytenoid tissue and that tissue will flop into the glottic inlet. There's floppiness to the epiglottis where it will actually fall posterior into the glottic area. Often the aryepiglottic folds are quite tight. Babies by definition usually have a more omega-shaped epiglottitis. So that in itself is not abnormal. It's the floppiness that you're looking for.

Dr. Alyssa Smith:

So is there any role for imaging?

Dr. Laura Ovidas:

I think in cases where the noise and the exam don't fit, you need to look further whether you get airway films, in other words looking at the subglottic area to make sure there's no narrowing, or tracheal airway

films which can tell you whether the trachea is collapsing, or an actual look in the operating room to determine whether there's any anatomic problems that you can't see at the bedside.

Dr. Alyssa Smith:

Should children either with or without dysphasia undergo a swallow evaluation with either a fiberoptic endoscopic evaluation of swallowing, FEES, or with a video swallow study?

Dr. Laura Ovidas:

I think they're both important and they are complementary. I don't think one versus the other is the best study. They both give you different information, but they're both good to determine whether the swallowing issues are clinically relevant, whether the child is aspirating. Some of the historical factors should help you determine that too. If they're sick all the time, if they've been in the hospital for pneumonia and things, you have to suspect desperation and they may require bronchoscopy and a BAL in the future.

Dr. Alyssa Smith:

Is there any role for polysomnography in workup of these patients?

Dr. Laura Ovidas:

Well I think many times we'll have parents that come in with video of their child sleeping, and if there's abnormalities there definitely, they should receive polysomnogram to rule out any true obstructive problems that could lead to cardiac issues or other issues down the road, but also may help document the need for a supraglottoplasty in a child with significant laryngomalacia.

Dr. Alyssa Smith:

And then are there any associated syndromes that we should be thinking about for patients with laryngomalacia?

Dr. Laura Ovidas:

Well certainly any syndromes that cause hypotonia. By definition, anything that's going to cause you to be floppier is going to give you increased risk of having things fall into the airway. I think you'll see that in some of the neuromuscular disorders and trisomy 21, those children probably need looked at a little bit more closely than your average healthy baby.

Dr. Alyssa Smith:

You mentioned previously taking patients with laryngomalacia to the operating room for microlaryngoscopy evaluation, but who should go to the OR for a more complete airway evaluation or does everyone require evaluation in the operating room?

Dr. Laura Ovidas:

Well I think, like I said earlier, that if you've got a child whose anatomy, when you scope them in the office or at the bedside, does not match with the symptoms that they're having, you need to make sure you're not missing something more distal, a distal airway lesion. Also, the children who are really having difficulty, cyanosis, failure to thrive, are not able to gain weight. Those kids, you really need to think about doing something surgically. Often, we'll start first with antireflux treatment. I've had children who,

with just antireflux treatment alone, have turned the corner in two weeks and have gotten significantly better, but it's the kids that respond to nothing and are continuing to have problems that you need to think about surgical intervention which would be a supraglottoplasty at this point.

Dr. Alyssa Smith:

So we've already started to dive a little bit into treatment approaches, but let's first start by discussing how you determine which patients can be managed conservatively and which patients may require surgical intervention to start.

Dr. Laura Ovidas:

Very few have I ever taken to the operating room immediately. I think that would be in the case of somebody that was critically ill. Most of the time we try something first whether it be watchful waiting, and that's the vast majority of the kids. I think a lot of kids, even their pediatricians know when they send them to us that it's likely just going to be, "Yeah, it looks like you have mild laryngomalacia. We're just going to have you treat this conservatively with positioning, with feeding, maybe shorter feeds." Sometimes we can send them to occupational therapy for some help with feeding, but the vast majority can just be monitored.

I usually see them a month to six weeks later, make sure they're still doing well, and usually they're even getting better. Occasionally, you'll have the kid that spits up a lot and often antireflux medications will help here. Unfortunately, we can no longer have the use of ranitidine. So we're usually talking about a proton pump inhibitor at this point. And then finally, there's the children that just don't get any better with medication or are severely affected by the laryngomalacia. Those are the ones that we usually take to the operating room and often find that surgical involvement really makes a difference.

Dr. Alyssa Smith:

You mentioned that occupational therapy can be helpful for speech and swallowing. What type of things do they generally try for these patients?

Dr. Laura Ovidas:

Well there's pacing, the way you feed, the amount you feed, and the biggest thing especially in the child that there's concerns for aspiration would be to thicken their feeds. There's some controversy as to when and what you can use in children to thicken, and I would leave that up to a discussion with your occupational therapist.

Dr. Alyssa Smith:

Moving on to surgical approach for these patients, you've mentioned supraglottoplasty, but can you expand on exactly what that entails?

Dr. Laura Ovidas:

Sure. I think the vast majority of the time we can get away with dividing the AE folds if they're tight. And again, this is something where you have to get a good idea of where the obstruction is coming from to determine what's the best operation to do. If it's just the epiglottitis flopping back into the airway, often it takes an epiglottopexy to actually tether that up so that it doesn't do that. If it's tight AE fold, sometimes you can get by with just releasing the AE folds, but often and I think most of the time there is

some element of arytenoid prolapse that you have to take off some of that redundant tissue. That can be done with laser, cold knife, microdebrider. I've done it in all three of those ways. I currently use the microdebrider.

Dr. Alyssa Smith:

And then how are these patients managed post-operatively?

Dr. Laura Ovidas:

It kind of depends again on the operation itself. How much did you do? How much concern is there for scarring? I used to leave children all intubated overnight when we did a bilateral procedure. I don't necessarily do that anymore if we're convinced that we did a good operation and there's no concerns that we're going to have any scar tissue formation. I usually will not feed them til the next day. If there was concerns for feeding preoperatively, we'd probably get a bedside swallow before we will consider sending them home and feeding them normally, but I've had children where we did a unilateral operation and it took care of their feeding from day one. So I would say the vast majority of children do stay in the hospital. I think anytime you're working on somebody's airway, it's probably prudent just to keep them at least overnight one night so that you monitor them and make sure that there isn't going to be significant swelling or other concerns that could make this a problem if they were at home.

Dr. Alyssa Smith:

What are some complications of supraglottoplasty that we should be aware of?

Dr. Laura Ovidas:

I've only once in my career seen a posterior glottic stenosis, a young lady, was not my patient. Just have to put that in there, but it's a very impressive picture. She does have some issues with exercise. We've talked about whether there was a possibility of releasing this scar in the posterior glottic area. She also has some asthma. So it's been very difficult to ascertain whether or not this is primarily caused by her glottic stenosis or it's from her asthma, but they've elected to currently just watch it. I see her about once a year. Other than that, you certainly have to be careful in the child who has feeding problems, especially aspiration, because you certainly don't want to make the feeding problems worse. Sometimes if you're taking off redundant tissue that's actually protecting their airway, you can end up with the aspiration actually being worse afterwards. So this has to really be investigated prior to consideration for operation in the first place.

Dr. Alyssa Smith:

Thinking about the preoperative visit that we have with these patients, how do you counsel parents on the chance of having a successful surgery?

Dr. Laura Ovidas:

I would say it's pretty high. I don't know that I have a specific number to say it's 90% successful, but I think based on what you see anatomically, I think that also tells you how much you're going to be able to help. For example, the child that has hypotonia everywhere, the likelihood that a supraglottoplasty alone is going to take care of their breathing issues is much lower than the otherwise healthy child who just happens to have a lot of redundant arytenoid tissue. That child you're probably going to do very

well with. So you almost have to base your guess on what you're seeing preoperatively, but you never can be 100%.

Dr. Alyssa Smith:

And so how do we define surgical success? Is it resolution of symptoms, improvement on objective testing?

Dr. Laura Ovidas:

I think it's primarily resolution of symptoms. If you've got the child that's undergoing failure to thrive because they can't get in enough calories because they're burning so many trying to breathe and now they're breathing significantly better and gaining weight, I would consider that a success, or the child, like I talked about, that was having horrible feeding issues, we did a unilateral supraglottoplasty. The kid started eating the next day and I never heard from them again and he did well. Those are successes. It's the child where you do the operation and there really is little to no improvement. That's difficult to say, was it the operation? Is that the child? Is it something else? And then you have to continue to investigate.

Dr. Alyssa Smith:

Thinking about followup, what does longterm followup look like for these patients?

Dr. Laura Ovidas:

I think we usually see them within a week or two after surgery just to make sure they're healing appropriately and everything looks well from that standpoint and then probably just once more after that to make sure that they're gaining weight, not noisy, parents are happy. If they're doing well, you can usually dismiss them because I rarely have had to do a second operation on an otherwise normal kid.

Dr. Alyssa Smith:

So this goes into the natural history of patients with laryngomalacia and thinking about some of the patients that we're treating conservatively or with watchful waiting, what is the timeline for potential spontaneous resolution?

Dr. Laura Ovidas:

I'm amazed how often some of them get better super fast. Some of them you'll see six weeks later and they're already better. I think a lot of that has to do with their eating performance and the fact that they are spitting up less, but in general, usually laryngomalacia with no treatment will resolve somewhere between 12 and 18 months of age.

Dr. Alyssa Smith:

For patients that don't have resolution of their laryngomalacia, what are some complications of untreated laryngomalacia?

Dr. Laura Ovidas:

Well I think anything that has a complication of sleep apnea can be a problem. It also can cause longterm problems with feeding. Children whose laryngomalacia is bad enough and it's causing

significant feeding problems where they're, one, not getting enough calories and two, having difficulty breathing when they do eat may require something like a G-tube. I think you probably need to do at least a sleep state endoscopy, if not a full polysomnogram in a child that you don't feel like you made any better. I also think you need to make sure that they are not aspirating via video swallow and/or feeds evaluation. Sometimes we don't fix them and they do need things like G-tubes and tracheostomies. I mean tracheostomy is the answer in a child that you cannot fix, and that is usually because they have multilevel airway obstruction, not just laryngomalacia.

Dr. Alyssa Smith:

Dr. Ovidas, thanks again for joining us. Is there anything else you'd like to add?

Dr. Laura Ovidas:

I don't think so. I think just be aware that the vast majority of these children are going to do just fine. I think especially first-time parents will often have a hard time with the fact that their kid sounds funny, but again, if the kid's growing, eating, happy, I think it's something they'll grow out of and you just need to counsel parents to not worry.

Dr. Alyssa Smith:

So in summary, laryngomalacia is a congenital softening of the tissues above the glottis that causes collapse and inspiratory stridor. It is the most common cause of noisy breathing in infants. In addition to stridor, patients can present with swallowing dysfunction, sleep disordered breathing, and failure to thrive. Nasopharyngoscopy is the gold standard for diagnosis and common findings include anterior prolapse of the arytenoid mucosa, short aryepiglottic folds, posterior collapse of the epiglottitis, and an omega-shaped epiglottitis.

Associated syndromes include down syndrome and any syndrome that can lead to hypotonia. Patients can be managed conservatively with watchful waiting, speech or swallow therapy, reflux medications, and many will have resolution of their symptoms by 12 to 18 months. Patients with more severe laryngomalacia or those that have failed medical therapy are good candidates for surgical intervention with supraglottoplasty. Supraglottoplasty is tailored to the anatomy of the patient and can involve division of the AE folds, removal of arytenoid tissue, or epiglottopexy.

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 should be on the differential diagnosis for infants with stridor?

For infants with stridor, our differential diagnosis should include laryngomalacia, subglottic stenosis, vocal fold immobility, laryngeal cyst, subglottic hemangioma, and vascular ring.

The second question is what are some common characteristics of stridor caused by laryngomalacia?

Patients with laryngomalacia will often have stridor that is inspiratory, worse after feedings, and worse when supine.

Finally, the last question is what is a complication of supraglottoplasty?

A complication of supraglottoplasty to be aware of is posterior glottic or supraglottic stenosis due to scarring.

Thanks for listening. And we'll see you next time.