Jason:

Hey there. It's Jason, one of the members of the ENT in a nutshell team. Thanks for listening to our program. If you enjoy it, please consider rating and reviewing the podcast and don't hesitate to contact us through headmirror.com with any questions or suggestions. Thanks and now under the episode.

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 otolaryngologists Dr. Karthik Balakrishnan. Today we'll be discussing pediatric vocal fold immobility. Thanks for being here Dr. Balakrishnan.

Dr. Karthik Balakrishnan:

Of course. Thank you Alyssa.

Dr. Alyssa Smith:

So when discussing this topic, we use some specific terminology and we're talking about vocal fold immobility. I think it's probably important that we start by a discussion of the terms immobility versus paralysis versus paresis.

Dr. Karthik Balakrishnan:

Yes. I think you're absolutely right. This is really important because terms have meaning and when things go into the medical record, they carry meaning forever. And so it's really important to be careful about the terminology we use. Immobility is probably the best term to use until you have evidence as to a potential cause when a vocal fold or both vocal folds aren't moving. Immobility is totally agnostic with regard to the cause. The words paralysis and paresis really imply a neurologic cause of some variety and it's important I think to include a fourth term as well of fixation, which would involve some mechanical prevention of focal fold movement whether Cricoarytenoid joint fixations, posterior glottic stenosis, scarring of the cord, something like that.

Dr. Alyssa Smith:

Now that we have that in mind, let's talk about presentation. And I think when we talk about presentation, it's important to break this down into unilateral immobility and then bilateral immobility. So focusing first on unilateral vocal fold immobility, how does a patient typically present?

Dr. Karthik Balakrishnan:

When we think about the three key functions of the larynx namely voice, breathing and swallow, that's usually the best way I think to break this down. These patients with unilateral vocal fold immobility are typically going to present with voice problems in terms of a weak cry, a breathy or hoarse voice. In terms of swallowing, they can have dysphagia and aspiration related symptoms as well and in some cases can actually have breathing related problems with stridor. That's going to be most common with the younger children and infants and it's sometimes positional where when the immobile cord is down, it falls out of the airway and opens the airway. Whereas when it's up, it falls into the airway and obstruct and you'd expect that for example with a flacid, paralyzed cord. Kids with bilateral vocal fold immobility present maybe a little bit differently though there is some overlap. Typically here, the main problem is with airway obstruction and voice and dysphasia or swallowing symptoms are a little bit more variable.

There has been a lot of discussion in the literature previously about the position of the vocal fold and whether that tells you anything. And it kind of does if the chords are more closed, then you're going to have more in terms of airway symptoms. And if they're more open, you'll have more voice and smaller problems, but there's quite a lot of overlap.

Dr. Alyssa Smith:

And then thinking about timing of presentation, are there typical ages that these patients will present that?

Dr. Karthik Balakrishnan:

Yes. For unilateral vocal fold motion, you can have a presentation at birth. More commonly, it presents after surgery and we'll talk a little bit later I think about common operative interventions that can lead to this. Up to 20% of a unilateral vocal fold immobility can be present at birth associated with forceps delivery as well. Bilateral vocal fold immobilities typically present at birth but it can present later with particular causes, particularly neurologic disease, Chari malformation, things like that. Most commonly is going to present that in birth.

Dr. Alyssa Smith:

And thinking about this settings that we can see pediatric patients, whether it be inpatient, on the floor or in the PICU or NICU versus the ED versus in the clinic. Is there a typical setting that these patients will present in?

Dr. Karthik Balakrishnan:

Yeah. And it tends to relate to the type of symptoms they have so the bilateral vocal fold immobility patients will often present in the NICU or shortly after birth in the hospital setting. The unilateral vocal fold immobility patients can present really at any of those settings depending on the cause

Dr. Alyssa Smith:

All right. So now that we've discussed presentation, let's move on to pathogenesis. And I think we all know that the mobility of the vocal folds is dependent on the function of the recurrent laryngeal nerve. And so, can you review with us the normal course of the recurrent laryngeal nerve?

Dr. Karthik Balakrishnan:

Sure. The recurrent laryngeal nerve has that name because it recurs, it makes essentially 180 degree turn so it comes as part of the vagus nerve, branches off from the vagus nerve in the upper chest and then on the left side, it loops under the aortic arch right next to the ligamentum arterial or ductus arteriosus depending on the age of the patient and then runs back up the neck in the tracheoesophageal groove typically until it enters the larynx just adjacent to the cricothyroid joint. On the right side, it follows a very similar course though instead of looping around the aorta, it loops around the innominate artery. It should be noted that you can't have a non-recurrent, recurrent laryngeal nerve that can be associated within aberrant retroesophageal subclavian artery. That's an unusual problem, but worth being aware of.

Dr. Alyssa Smith:

With that normal course in mind, what are some causes of vocal cord immobility?

Dr. Karthik Balakrishnan:

Well, as you say, the anatomic course of the nerve has a lot to do with that. So unilateral, you can have birth trauma. The causes of that are a bit debatable but some people will talk about traction injury to the neck or even compression. There has been one study actually looking at birth trauma versus truly idiopathic congenital which is kind of an interesting study. And while it was a little difficult to differentiate the two, it's suggested that the birth trauma patients were more likely to have unilateral immobility and more likely to recover spontaneously. You can have direct laryngeal trauma from suctioning, traumatic intubation, things like that. You can have thoracic disease processes like tumors. I mean, in adults, you hear stories about things like ascending aortic aneurysms and things stretching the nerve.

You don't really see that as often in kids. Certainly, thoracic and cardiothoracic surgery can do it. The most common one there's probably PD ligation happening in about 1 to 7% of cases, but really any aortic arch surgery puts that at risk. You can have of course, thyroid surgery and then a central or peripheral neurologic disease. Interestingly in kids, a lot of these are idiopathic as well we don't really know why. And then really any surgery in the tracheoesophageal groove. So tracheoesophageal fistula repair, branchial cleft anomalies that run nearby, things like that. With bilateral immobility, the most common thing we think of in kids is Chari malformation and that can be a result of essentially the posterior fossa contents herniate down and that causes pressure on the vagus nerves as they exit the skull base. But that can also be an intermittent thing where the carrier can kind of drop down and cause compression and then rise back up and you can have intermittent symptoms.

Meningomyelocele can do it and then more kind of chronic neurologic disease, spinal muscular atrophy, congenital myasthenia gravis, head bleeds can do it and again, some of these can be idiopathic. And interestingly, there's actually a few reports of heritable, familial bilateral vocal fold immobility that can be either autosomal dominant or X linked and seems to be associated with digit abnormalities.

Dr. Alyssa Smith:

All right. So let's move on to workup for these patients. What are some important history questions that we should be asking?

Dr. Karthik Balakrishnan:

As with any pediatric airway question, a lot of the questions or complaints a lot of the questions will overlap. So we want to about the birth history, how was the child delivered? Were they term or not? We want to know about whether they required any respiratory support at birth, whether intubation, oxygen CPAP, we want to know when the symptoms started, how soon after birth. And then we want to know what's the impact on the kid? Do they have other respiratory symptoms, cyanosis, retraction apnea, have they had breweries or needed CPR? Have they had any previous airway surgery such as needing a tracheostomy for instance or interventions for their vocal folds? Have they had any other operations? Cardiac surgery, thoracic surgery, TEF repair like we discussed. We want to know about their neurologic history. How's their tone and develop and otherwise? And then that's leads into things like growth history, motor development.

Then we want to know about the other functions of the larynx. We want to know about their voice, what's their cry like? Or if they're old enough to speak, what's their voice like? Do they have vocal fatigue? Do they lose their voice when they try to yell or cry because they can't maintain a good closure of the glottis. And then we want to know about swallowing. Do they have dysphagia symptoms? What are they eating now and how are they getting it? Bottle versus breastfeeding for example, what textures do they take? Do they have any signs of aspiration, recurrent respiratory infections? Have they had

previous swallow studies and what did those show? Those would be some common things we'd want to know. And then we want to have a broad differential diagnosis for airway related symptoms so thinking about other potential causes of similar symptoms is important as well and then asking relevant questions there.

Dr. Alyssa Smith:

And so moving on to our physical exam, what are some specific physical exam findings that we should be looking for?

Dr. Karthik Balakrishnan:

Yeah. Externally, step one is again, assess your ABC's. Does the child look stable? Are they in acute distress? If you have the ability to do vitals in the clinic or in the setting where you're evaluating the child, then that's helpful. Saturations, respiratory rate, heart rate is important. Then in addition to a full head and neck exam, looking for other congenital anomalies, things like that, you want to do your good airway and respiratory exams. Do they have retractions, cyanosis, accessory muscle use increased work of breathing in any way? Do they have stridor? If so, is it inspiratory versus excretory or is it biphasic? What's their voice and cry like? If they don't cry during the exam, they'll probably cry later when you do your flexible laryngoscopy. You want to listen to their lungs but then you also want do careful auscultation of the heart because again, a lot of these kids will have congenital heart disease or something else and maybe they've had cardiac surgery, maybe they haven't.

But as the clinician, that's part of your responsibility to examine as well. The flexible laryngoscopy is I think essential part of the physical exam here as well. And so being prepared to do that is important. It can be tricky in a young baby but getting a good view of that glottis to really assess motion is important. There are some data that suggests that inter-rater reliability for flexible laryngoscopy and vocal fold motion is great. If you say, yes there's motion or no there's not, but the subtleties of is it normal or is it weak are not quite as reliable. Having a sort of consistent ability to judge in your own mind, I think is important.

Dr. Alyssa Smith:

And then with those physical exam and laryngoscopy findings in mind, I think that there's a few other investigational tools that are available. And so can we first start by discussing the use of ultrasound in diagnosis?

Dr. Karthik Balakrishnan:

Yes, of course. There are several people in the United States and elsewhere who have done a lot of great work on ultrasound for laryngeal assessment in children. There's some data that suggests that you can assess the larynx effectively with ultrasound even into the third and fourth decade of life before things get too calcified. So the ultrasound, I typically prefer to use the small hockey-stick probe with the SonoSite ultrasound, but you can use whatever you have. A lot of people use small portable tablet based ultrasounds as well. It is fine if the baby's crying, but keep in mind that if they are crying, young babies in particular will maybe not snap their vocal folds open as much when they take a breath in during crying. Typically, I just use a transverse or axial view and identify the air column of the trachea and then scan upwards until I can see the arytenoid and glottis and then I just sit there and watch for a while.

One thing I would say, it's important to try to keep the ultrasound plain, very flat, very axial, because it's easy to tilt it when you get under the pudgy chin of the baby. And then it's very hard to interpret I think at that point. It's also worth recording it so you can look at it later and confirm. I will say

that ultrasound is not perfect. It is generally pretty easy to interpret if you know what you're looking for but sometimes it will disagree with flexible laryngoscopy and if that's the case, then I would tend to trust the flexible laryngoscopy more.

Dr. Alyssa Smith:

And then how about laryngeal EMG? Is that used in the pediatric population?

Dr. Karthik Balakrishnan:

It is used, it can be used and can sometimes be very useful. Laryngeal EMG I think has use in a couple of applications. The first is it's one way to differentiate a neurologic etiology from a fixation or if you're not sure both exist, it's a way to look for that. Another is to look for recovery of function as with any other EMG. The challenge in kids unlike adults is that most kids are not going to put up with EMG needles being inserted transcervically when they're awake and then doing voluntary voicing and so on. What typically ends up happening is this has done under a light general anesthetic, a similar plan to a sleep state endoscopy maybe or a little deeper. And you can either put the needle electrodes transcervically or some people have actually put them transorally with a wire attached to something like a butterfly needle and inserted them into the vocalis muscles that way. I typically do it transcervically. And then with the child under light anesthetic, hopefully they're breathing spontaneously enough that you can actually see the abductors in action when they take an inspiratory breath.

Dr. Alyssa Smith:

And then how about the role of video swallow studies?

Dr. Karthik Balakrishnan:

Yeah. Video swallow studies and other FEES, these instrumentals follow studies are important because many of these children as we discussed can have swallowing symptoms, and how it looks on flexible laryngoscopy in terms of the position of the cords and so one does not necessarily predict what their swallow function is like and their airway protection is like. I tend to do at least one of those two studies on everybody, unless they're an older kid who has a unilateral immobility who has zero symptoms whatsoever, who's neurologically otherwise normal and it's sort of an isolated problem of unilateral vocal fold immobility, maybe I'd skip it on them. But the thing is, especially in the young kids, if you're putting a flexscope in the nose anyway, it takes very little additional effort to just try to do a FEES study and see what's going on.

Another option is what's called a SEES study, Static Endoscopic Evaluation and Swelling, where you feed the child a little bit of green dyed material and then put the scope in and just see if there's anything left or anything obviously in the airway. That can be a good backup option as well. And these are all useful studies to do potentially before you go to the operating room to do a more formal airway endoscopy.

Dr. Alyssa Smith:

And then are there any other imaging studies that could be helpful in this setting?

Dr. Karthik Balakrishnan:

Yeah, definitely. It really depends on your suspicion for certain causes. Certainly if you have a child with bilateral immobility where you suspect a neurologic cause or you don't have another obvious

explanation, then MRI of the brainstem is very helpful to evaluate [inaudible 00:16:48]. Again, sometimes that [inaudible 00:16:50] can be intermittent so I have had a patient in the past where the initial MRI didn't catch it and the child had intermittent symptoms and essentially a CT MRI had to be done to capture that. If you think there's thoracic pathology then CT or echo or whatever else depending on your suspicion maybe appropriate as well. And then sometimes you just do a full MRI of the course of the recurrent laryngeal nerve if you can't find any etiology. I would say from experience, that's not always particularly revealing.

Dr. Alyssa Smith:

And then before we move on to natural history, are there any associated syndromes that we should be thinking about for these patients?

Dr. Karthik Balakrishnan:

Yeah. There are a few. Moebius syndrome is one that's very much worth mentioning. People usually think of facial nerve problems with Moebius syndrome, but I believe that the vocal fold mobilities is a very common, I think it's the second most common cranial nerve affected. And then we talked a little bit about how some of these kids can have bilateral familial immobility. And then I think really anything where a kid has a history of something that might lead to increased risk surgically for recurrent nerve injury for example charge, VACTERL anything where there's a TEF for instance, those kids are going to be more prone to needing surgery and therefore potentially have vocal fold immobility from a recurrent nerve injury. And then those things are worth considering as well if you're going to take the kid to the operating room for an MDLB because they might affect operative risks or they might lead you to look for other airway things as well.

Dr. Alyssa Smith:

You mentioned microlaryngoscopy and bronchoscopy as a method for a continued workup of these patients. Can you speak a little bit about that?

Dr. Karthik Balakrishnan:

Yeah. MLB or MDLB is an important tool in the assessment of these patients often combined with flexible bronchoscopy. Microlaryngoscopy and bronchoscopy lets you do a few things. The first thing it does and it lets you get a really detailed anatomic look at the chords and the glottis as well as the rest of the larynx. Another thing it lets you do is to palpate the cricoarytenoid joints to see if there's any fixation. The MDLB also lets you look for things like posterior glottic stenosis that may tether the vocal folds and maybe tricky to see a on flexible laryngoscopy in the awake child. And then it lets you look for other associated airway abnormalities such as TE fistula, laryngeal cleft or other things that may affect your clinical decision making. The flexible bronchoscopy helps you look at the lower airways, look for sequelae of aspiration, look for dynamic vocal fold problems or other airway problems that might not be as evident on rigid endoscopy. I think if I have any question in my mind on why a child has a particular vocal fold immobility concern, I would have a very low threshold to do operative airway endoscopy.

Dr. Alyssa Smith:

All right. Moving on to natural history, how many patients will just recover spontaneously from a vocal fold immobility?

Dr. Karthik Balakrishnan:

The good news is that kids tend to recover. About 70% of non-iatrogenic unilateral vocal fold process will resolve spontaneously. Again, if it's a birth trauma, maybe that recovers a little bit more than an idiopathic congenital unilateral immobility. Typically recovery is going to occur in the first six months. When we look at kids who have had cardiac surgery, the data such as that about a third or 35% or so will recover spontaneously and if you have bilateral vocal fold immobility, about 65% of those will recover over time. It's not always a symmetric recovery. You can have partial recovery on one side and complete recovery on the other for example and the time course is very variable. In general, if there are no associated abnormalities, the prognosis is better. Most people would argue that the recovery is likely to occur within the first two years, maybe the first three years, and that of the recovery occurs after three years or so it may be incomplete because the laryngeal muscles are atrophied.

You may have cricoarytenoid fixation or maybe you'll have synkinesis because there's been some reinnervation that's not entirely normal. There are some people who argue that in these patients, you can have recovery several years later and so you should not do aggressive destructive intervention for that. I think that that opinion is currently in the minority, but it may change over time.

Dr. Alyssa Smith:

Okay. Let's talk about treatment now. In general, what is the treatment approach for these patients and given that so many of them may recover spontaneously, can these patients be managed conservatively?

Dr. Karthik Balakrishnan:

Definitely. The decision to treat really depends on the individual patient and what are the consequences they're having from their immobility, how is it affecting their function and what are their other comorbidities? Just to give you a quick example, if you have a child with a unilateral vocal fold immobility after aortic arch surgery, their chance of recovery again is pretty good and maybe a third will recover and they recover pretty promptly. But if that child is aspirating floridly, you may be very concerned about soiling the lungs in a child with medical fragility and complex congenital heart disease. You may be more prone to intervene despite their decent chance of recovering. That said, if a child is executing their key laryngeal functions of breathing, swallow and voice effectively and staying healthy, you can manage them conservatively even if their laryngoscopy looks terrible. In the very young kids, voice is probably the least concerning. Airway first, then feeding, them voice and the older kids that order may be rearranged.

Dr. Alyssa Smith:

And so thinking about timing as far as who would be a good candidate for surgical intervention, is there a specific time that you would allow for spontaneous recovery before you would proceed with maybe permanent intervention?

Dr. Karthik Balakrishnan:

Yes, there is. I would wait to do anything destructive or irreversible or anything that might affect the ability for the child to recover spontaneously. I would hold that off for at least 18 to 24 months if I can.

Dr. Alyssa Smith:

And then thinking about the variety of surgical options that are available, let's first focus on a unilateral vocal fold immobility. What are the different surgical options that are available for this patient?

Dr. Karthik Balakrishnan:

The surgical options in kids are very similar to those in adults, but we weigh them a little bit differently. So again, in unilateral paralysis or immobility, the problem is that there's typically too much of a glottic gap leading to trouble with voice and swallowing and potentially difficulty with the airway. One option that's commonly used is injection laryngoplasty essentially bulking the cord and medializing it a little bit. This is typically done under general anesthesia unlike adults you typically can't get away with awake injection in kids. The procedures done with microdirect laryngoscopy and bronchoscopy, palpating the cricoarytenoid joints to rule out fixation, ruling out any other potential etiologies. And once you see that things are good, then you can inject that material into the paraglottic space. I typically will do two sites of injection. One, just enter lateral to the vocal process through the arcuate line to try to get the vocal processes rotated immediately in the posterior cord medialized.

And then one about a third of the way back from the anterior commissure to get the anterior cord medialized. There are multiple types of materials that can be used and people debate which are safe and appropriate. Common ones that are used include calcium hydroxyapatite which lasts for an undetermined period of time but may be permanent. Some people will use the carrier for that which the brand name for that is a Prolaryn gel which lasts four to six weeks as a temporary intervention. Some people will use cadaveric dermis or Symetra. You can use gelatin powder, gelatin sponge like gelfoam and you can use autologous fat. Those would be kind of the common ones that are used. You have to be careful when you inject because if you over-inject a child, you can significantly narrow the airway and cause airway compromise.

It doesn't take much in a baby. The benefit lasts variably. The thought is, and this is extrapolated in some ways from the adult literature, is that the benefit can last longer than the injected material is present because if you reposition the cord into a better place and then the person develops some synkinesis in that position, they'll end up better off in the long-term. Some people do need repeat injections though as the material fades. Another option would be medialization laryngoplasty thyroplasty. I don't tend to do that very often in kids and I think most people do not for a few reasons. The first is, unlike adults again, most kids are not going to put up with voice tuning interrupt with a flexscope in the nose and them half awake and you manipulating the implant.

Another part of it is that as children grow, you may have to replace the implant and I have heard some laryngologists say that they worry about implanting very young people because there's just more time for the implant to extrude or have complications as well. I tend not to favor that procedure in young kids. The combination of thyroplasty and arytenoid abduction is also possible. It's been reported in kids again, not done very commonly. The more preferred option in kids who have gotten to the point where they're unlikely to recover, in other words it's been 18 to 24 months, would be laryngeal reinnervation. There is a study from the laryngoscope from several years ago demonstrating that in young adults even up through early middle age, that reinnervation tends to have better outcomes than thyroplasty. And certainly in kids where their plasticity is so much more, I would tend to believe that even more.

There are lots of different ways to do reinnervation. The ansa cervicalis recurrent nerve is the most common for unilateral immobility in children when it's from a neurologic source. This restores vocal fold tone but it does not restore motion. Remember, you're essentially randomly reinnervating the motor neurons and therefore you're likely to get synkinesis. And the idea then is you want to get synkinesis in a better position so this is often combined with injection laryngoplasty. Usually start to see results from the reinnervation in three to six months, but you'll see them sooner if you do the laryngoplasty injection at the same time. And there's some data from Karen Zur at CHOP for example, that this can also be helpful. Reinnervation can also be helpful for kids who have dysphagia and aspiration problems related to unilateral vocal fold immobility from a neurologic cause.

Dr. Alyssa Smith:

And then how about some of the options available for a patient with bilateral immobility?

Dr. Karthik Balakrishnan:

For bilateral immobility, the tables are turned a little bit and the priorities are a bit different. So these kids again, typically will have airway symptoms as their biggest problem and voice and swallow as perhaps secondary problems. One option is simply to do tracheostomy and that is sometimes what you end up doing. And there's some people who favor that because it's non-destructive in a lot of ways, it does not affect the chance for spontaneous recovery. And maybe it'll take several years but these kids may recover function enough or grow their airway enough that you can decannulate them without other interventions. That said, I think we're all aware that tracheostomy in young children has its own significant drawbacks which we're not going to go into that here so what are the other options? There are a lot of other options. The nondestructive options here include temporary suture lateralization.

I think that is truly a temporary procedure in children perhaps unlike adults, the suture typically cheesewires through the cord and the cord goes back to where it was fairly rapidly, but it is theoretically irreversible though you can have scarring laterally of the cord with it. You could do more destructive things like cordotomy or cordotomy plus partial arytenoidectomy. If I'm going to do that, I'll often combine that with a suture lateralization so when you release the cord from the vocal process and it becomes thick and retracted anteriorly, I'll then lateralize that anterior remnant so that it scars out and creates more space. You can do an arytenoidopexy as well. Oftentimes when you have a neurologic origin, you have a flacid cord and arytenoidal tip forward immediately and so you can tip it back and laterally or you could do somewhat less destructive things.

For instance anterior posterior cricoid split, which can be done endoscopically in the vast majority of kids. You lead them intubated with a large tube for about 10 days and there's at least one study that suggests that you can have about a 75% success rate with that in terms of either avoiding tracheostomy or decannuling children who are already tracheostomized. I'm not sure the success rate is that high now that we've had further experience, but it's a great option to avoid tracheostomy if you've tried everything else.

You can do an endoscopic posterior cricoid split with a cartilage graft so an endoscopic posterior Laryngo-Tracheo-Plasty. That's a great option. I would typically reserve that for kids who are six months to a year or older but we have done it younger and succeeded. And then there is a new option which is the phrenic to recurrent reinnervation which has been done in adults. But there are a few practitioners including one currently in the United States who are doing this for children and the early results seem promising. I think this is a technically very demanding operation and it's something that needs more study in kids, but may be become a very viable option for appropriately chosen patients with bilateral cord immobility in the future.

Dr. Alyssa Smith:

And then thinking about management postoperatively, how are these patients typically managed? And I think we can probably break this up as well in the unilateral and bilateral groups.

Dr. Karthik Balakrishnan:

Unilateral, again it depends on what their symptoms are. Oftentimes, if voice is the main concern, then you focus on that and then you follow them up especially the older kids with things like voice therapy, videostroboscopy as you would with an adult patient. If dysphasia is the problem, then you may get a follow-up swallow study and track them that way. It really is driven a lot by their symptoms

preoperatively. The bilateral vocal fold immobility kids can be a little more involved again, because you're focused mostly on airway. And then it really depends on what your intervention was. If you did a tracheostomy, then I would follow them in clinic or in the hospital as I would any other tracheostomy patient but I would tend to flex scope them every time in clinic to see if they're recovering any function. And I think really for either unilateral or bilateral all other things being equal, I would want to see these kids in clinic every three to six months to do a flex scope and a stroboscopy if they're old enough to assess whether they've had recovery of function.

Dr. Alyssa Smith:

And then I think an important part of this is counseling the parents or the team taking care of the patient on outcomes. So first focusing on if a patient is aspirating pre-op, what can we tell the team or the parents about whether an injection or other intervention will help?

Dr. Karthik Balakrishnan:

That's a really important question. Counseling is critical here because none of these things is a guarantee. And as with any other laryngeal intervention, improving one of the laryngeal functions may compromise some of the others. For instance, if you do an injection laryngoplasty for a kid with unilateral vocal cord immobility who has some dysphagia aspiration problems, that may be very, very successful and there's some data that suggests that about two thirds of patients can advance their diet after injection and that's even very young kids. But that's two thirds of kids so that means one third maybe won't be able to. It's important to set realistic expectations and to make it very clear to families that the child will need ongoing follow-up particularly as the injection material fades.

Dr. Alyssa Smith:

Thinking about a patient that may have difficulty with their voice, another function of the larynx, how can we counsel parents or the patient even on improvements that we can expect in their voice?

Dr. Karthik Balakrishnan:

It's a little easier to be confident about voice I think than it is to be confident about swallowing function after these interventions. If a kid has bilateral vocal fold immobility and significant airway problems, then I would tend to counsel the parents that the voice is probably going to be worse after surgery, but often children compensate. And so the voice may get worse for a while and then improve somewhat though it may not be totally normal. And so that's an important discussion to have with parents preoperatively is whether that's a trade-off they're willing to make, most parents are. With unilateral immobility, it's a little more established probably because we've learned a lot from our adult colleagues that injection medialization, reinnervation, they all tend to be quite successful. They often will require voice therapy and I usually will send kids to voice therapy beforehand if they're old enough so that they're prepped and ready to go.

Dr. Alyssa Smith:

And then moving on to decannulation, for a patient that has bilateral immobility and needed a trach, how do you counsel parents about the chance of decannulation in the future?

Dr. Karthik Balakrishnan:

I would tell parents in that case that the chance of decannulation is excellent but it really depends on what interventions you have to do in the meantime. Again, if we watch and wait, a reasonable number

of these patients will recover over time. And that's assuming a neurologic cause, if the child has a Chari, that Chari is repaired, then the chance of improvement is very good. If there's scarring stenosis fixation, then we may have to do airway reconstruction, so it really on the cause and what interventions are required. But assuming that the child has an isolated glottic obstruction due to bilateral immobility, that's pretty good candidate to do something to get that trach out.

Dr. Alyssa Smith:

And then thinking about the child that is now two or three years out from their diagnosis, we're pretty sure that they have a permanent paralysis. What do you tend to see as far as requiring additional surgery throughout their life?

Dr. Karthik Balakrishnan:

Again, it really depends on what we've done to them to begin with. In a kid who's had a trach, I would counsel the parents that especially if it's been a long-term trach that there's a decent chance we'd have to do something reconstructive to get the tracheostomy out. Whether it is to open the glottis more or whether it has to deal with suprastomal collapse for instance, we're likely to have to do something. If the child has already had let's say a reconstruction, then they're going to need ongoing follow-up to make sure that their airway stays patent and grows with them.

That said, as that kid gets older, we can do more and more of this in the clinic including things like awake tracheoscopy to examine the airway. If the child has had something to spread the posterior glottis like an anterior posterior split or a posterior graft, then that's a kid too where I would counsel the family and the child that when the child hits their teenage years, we'll want to watch their voice in particular carefully because there are some kids who then need a posterior cricoid reduction to improve their voice. But again, that requires long-term follow-up and individualized decision making.

Dr. Alyssa Smith:

We've talked about long-term follow-up schedule and what that typically looks like for patients, but are there any long-term complications that we should be looking for that are commonly seen or rarely seen in these patients?

Dr. Karthik Balakrishnan:

Definitely. So it goes back to those three functions of the larynx. If we do something to enlarge the glottic airway, then we have to watch for long-term consequences of dysphagia and aspiration and we have to watch for long-term consequences to the voice. And again, there are some people who show up years later in their teenage years or young adult years and they're not satisfied with their voice anymore because now their voice is more important to them either for work functions, for school, for social interaction and so on. Similarly, if there's any concern about glottic obstruction from an airway standpoint, then as those kids get older and they get into competitive sports or other activities that require good airway function or optimal airway function, things may manifest that weren't obvious to begin with. From an aspiration or dysphasia standpoint, we obviously want to follow them there because if they have long-term ongoing silent aspiration in particular that just doesn't get detected, that could cause significant lung damage. It depends on where they started and what's been done to them as to what the long-term complications might be.

Dr. Alyssa Smith:

All right. Dr.Balakrishnan, is there anything else you'd like to add?

Dr. Karthik Balakrishnan:

The only other thing I would like to add is I think as with any complex airway voice swallow concern, these children can get good care in a lot of places and it may be worth considering having them evaluated in a multidisciplinary aerodigestive or airway program so that all those different functions of the larynx and any associated syndromes or diseases or conditions can be assessed holistically and dealt with together because I think that that does the best service to the patient rather than focusing purely on voice or purely on airway for example.

Dr. Alyssa Smith:

All right. Dr. Balakrishnan thanks again for joining us.

Dr. Karthik Balakrishnan:

Thank you, Alyssa. This has been a pleasure.

Dr. Alyssa Smith:

In summary, when thinking about patients with vocal fold immobility and how they present, it's important to think about the three main functions of the larynx. These include voice, breathing and swallowing. Patients with a unilateral vocal fold immobility can typically present with stridor, a weak cry, a hoarse voice, and then dysphasia or even aspiration. A patient with bilateral vocal fold immobility will typically have symptoms more related to airway obstruction. Some common causes of a unilateral immobility include birth trauma which can be from a traction type injury from forceps delivery or even direct laryngeal trauma from traumatic intubation or aggressive suctioning. Can also be from a thoracic disease process such as a tumor, iatrogenic damage during thoracic or cervical surgery, and then a central or peripheral neurologic disease. It can also be completely idiopathic as well. A bilateral immobility can be caused by a Chari malformation, Meningomyelocele, spinal muscular atrophy, congenital myasthenia gravis and then subdural hemorrhage or other intracranial bleed.

We can also see bilateral familial vocal folds immobility which is associated with digit abnormalities. Some important history questions that we should be asking include a birth history such as whether or not there is any intubation or any airway difficulty at the time of birth, the age at which symptoms developed if they weren't present immediately at birth, their growth history, current diet including signs of aspiration, their surgical history specifically focusing on thoracic, thyroid and cardiac surgeries and then their overall tone and neurologic symptoms. When performing a physical exam, it's important to determine whether or not the patient is stable or unstable and perform our typical ABCs. We should also be assessing their voice and auscultating their lungs and their heart. Cardiac auscultation is important because a lot of these patients will present with cardiac abnormalities as well.

Flexible laryngoscopy is helpful for determining whether or not an immobility is present although it's not always completely sensitive for determining whether or not there is impaired mobility. Microdirect laryngoscopy and bronchoscopy under anesthesia can also help rule out any other airway anomalies, a cricoarytenoid joint fixation and then posterior glottic stenosis as well. Ultrasound and laryngeal EMG can also provide helpful diagnostic information and an MRI can be helpful to diagnose a Chari malformation or even evaluate the complete course of the recurrent laryngeal nerve to determine if there's any lesions.

When thinking about treatment options for a patient with a unilateral immobility, this is often a problem where there's too much gap in the glottis and therefore voice and swallowing problems. Some options include injection laryngoplasty for which there is a variety of materials that lasts a various amount of time and then laryngeal reinnervation, which as a reminder restores vocal fold tone but not

motion. Some older children may be a candidate for medialization laryngoplasty or thyroplasty but this is not done very commonly. For patients with the bilateral immobility, again, this is a problem of not enough glottic gap. These patients can sometimes require a tracheostomy. Some other interventions include suture lateralization of the vocal fold, a lateral cordotomy, arytenoidectomy, arytenoidopexy, anterior, posterior cricoid split and endoscopic posterior cricoid split with a cartilage graft.

Some of us [inaudible 00:43:14] are also looking at the possibility of reinnervation for these patients. Regardless of whether there's unilateral or bilateral immobility, these patients require long-term close follow-up and this is to assess for any complications such as glottic obstruction, dysphasia, silent aspiration, or worsening vocal outcomes. And as with most complex pediatric problems, these patients can be best managed by a multidisciplinary team. 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 typical course of the recurrent laryngeal nerve?

The recurrent laryngeal nerve is a branch of the vagus nerve and supplies all of the intrinsic muscles of the larynx with the exception of the cricothyroid muscles. So both the left and right recurrent laryngeal nerve exit the skull base as the vagus nerve, the left recurrent laryngeal nerve loops under the aortic arch next to the ligamentum arteriosum and the right loops under the innominate artery. They both then travel superiorly in the tracheoesophageal groove and enter the larynx adjacent to the Cricothyroid joint. The course of this nerve is important to keep in mind because any lesion along its course can result in a vocal fold immobility. The second question is what is the natural history of patients with a vocal fold immobility and how frequently can we expect patients to recover spontaneously?

Patients with a non-iatrogenic unilateral vocal fold immobility will typically recover spontaneously about 70% of the time. And this usually occurs in the first six months. For patients that have cardiac surgery and have iatrogenic injury, about 35% of them will recover spontaneously. And then finally for patients born with a bilateral vocal fold immobility, about 65% of them will have recovery of function. It's important to keep in mind that recovery is not always symmetric, that their prognosis is better if there's no associated anomalies and that if recovery occurs after about three years, it's usually incomplete and this is due to a laryngeal muscle atrophy, cricoarytenoid fixation and even synkinesis Thanks so much for listening and we'll see you next time.