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. Karthik Balakrishnan.

On today's episode we'll be discussing pediatric tracheal stenosis. Thanks for being here Dr. Karthik Balakrishnan.

Dr. Karthik Balakrishnan:

Thank you Alyssa.

Dr. Alyssa Smith:

So, to start, I know that these patients can present in a number of different settings. Whether it be in clinic or on the inpatient side, but how do patients typically present when they have tracheal stenosis?

Dr. Karthik Balakrishnan:

It's a great question. It can really vary quite a lot. One of the things that Dr. Wood at Cincinnati taught all of us was that you can't trust a baby because children can hide bad airway problems very effectively. Often because they just, for instance, don't take a deep enough breath to muster up enough airflow to generate stridor. So, really it can vary. But in general I think you could say comfortably that the symptoms vary depending on the severity, location and length of the stenosis. And also on the age and size of the baby and any associated health problems they might have.

Dr. Alyssa Smith:

So, when we get a consult on the inpatient's side for a pediatric patient with noisy breathing, what clues should we pay attention to that may make us have a high suspicion for tracheal involvement of stenosis?

Dr. Karthik Balakrishnan:

It's a really important question because in babies in particular, obstruction at one level of the airway can mimic obstruction at other levels of the airway very effectively. So, sometimes the presentation is not specific to a particular condition and it's more a question of being alert to the possibility of tracheal stenosis or other significant airway problems.

So, similar things we look at, for instance stridor. That's the classic symptom of noisy breathing related to turbulent airflow through a narrow or irregular airway. Generally, the stridor can be associated with the location in the airway. So, if it's a tracheal problem, particularly mid or distal trachea, you can have expiratory noise. If it's a cervical tracheal problem, you can have inspiratory noise. But those things are not entirely accurate. They're good clues but they're not perfect.

Otherwise, things like wheezing, a barky or harsh or brassy cough can be concerning. Retractions or increased work of breathing are very concerning. In fact, more concerning than stridor is.

Recurrent pulmonary infection, cyanotic episodes, all these are now called brui. These sort of dying spells babies can have, feeding difficulties, failure to thrive. In older kids you can see exercise intolerance. And then depending on how bad the stenosis is you can even have voice problems. If it either extends up toward the glottis or if the airflow is so poor that the child can't generate a strong voice.

Dr. Alyssa Smith:

So, before we move on to further work up of these patients, I think it's important to touch on a differential diagnosis and possible etiologies for this stenosis. Can you elaborate on things that we should be thinking about as far as different causes?

Dr. Karthik Balakrishnan:

Absolutely. So, we can think of three types of causes for narrowing of the airway.

The first is what you can think of as a true stenosis. An endoluminal stenosis or scar, that's going to come commonly from post intubation injury, congenital tracheal webs, thermal injuries, infection. Sometimes it can be entirely idiopathic and you never really know what caused it.

Then you can have more problems of the exoskeleton of the airway, cartilaginous problems. So things like complete tracheal rings. Which we can talk about a little bit later but essentially the normal horse shoe shaped cartilages of the trachea are replaced by complete O-shaped rings.

Or even a sleeve trachea, which is just a two-hoof cartilage. And then you can have more dynamic problems of the cartilage. So things like tracheomalacia. And then you can have extrinsic compression, which is not in and of its self a stenosis, but it can act very much the same way and that could be from things like vascular compression, most commonly. Mediastinal masses can do it.

Or you can have actually posterior compression from the esophagus. So, children who have a distal esophageal narrowing or obstruction or poor function can actually have impacted food... Cause posterior intrusion in to the trachea as well.

Dr. Alyssa Smith:

So, it looks like there's a lot of different things that we should be thinking about. And maybe we can go through a couple of them to touch on pathogenesis a little bit. So, as far as tracheomalacia goes, how exactly does that happen? What causes that increased compliance?

Dr. Karthik Balakrishnan:

So, tracheomalacia is different in children and in adults. So, in adults we're used to hearing this sort of post intubation tracheomalacia, where the ET tube cuff or trach cuff was blown up too high and you get ischemia of the tracheal cartilage and it becomes soft or floppy. Less common in kids.

And what we see more common in kids is either intrinsic weakness or softness or immaturity of the cartilage, or more likely a wide cartilaginous arch. So, instead of having your normal four or five to one relationship of cartilage to membranous trachea in terms of the circumference, it might be as bad as one to one, because you have a very large cartilaginous arch, and therefor a wide posterior membranous trachea as well.

And so then what ends up happening is, if it's a more distal tracheal problem or intrathoracic tracheal problem, when you breathe out, during expiration the intrathoracic pressure is higher than the intraluminal tracheal pressure and you get collapse.

You can have the opposite if its cervical involvement where when you breathe in you get collapse as well.

Dr. Alyssa Smith:

And then you also mentioned external compressions from things like vasculature, what are the different vessels that can cause compression?

Dr. Karthik Balakrishnan:

I think it's important first to distinguish tracheomalacia from vascular compression. A lot of people will do an endoscopy and see some, for example, innominate artery compression of the anterior trachea and they'll call it tracheomalacia. That's not tracheomalacia.

Tracheomalacia is collapsibility of the airway that varies with the repository cycle. Now that said, vascular compression can exacerbate tracheomalacia and so the most common thing was see in innominate compression, where the innominate artery runs superolaterally from left to right across the front of the mid-trachea and squashes the trachea a little bit. And we know that airways that are already a little narrow tend to collapse more and so that tends to be a collapse point for children who have a somewhat malacic airway to begin with.

Other common causes are what you might call rings and slings. So, vascular rings, double aortic arch is the most common. Typically, a dominant right aorta with smaller left aortic arch. And this happens in embryology when the distal right fourth branchial arch involute as it ought to and so you get this paired aortas.

And then you can see things like pulmonary artery sling, where instead of having symmetric pulmonary arteries coming off the main pulmonary artery, you have a very dominant right sided pulmonary artery and a small left one that branches off the right side and hooks between the trachea and the esophagus and causes some compression.

Dr. Alyssa Smith:

And then we've talked about lots of different things that can cause narrowing or stenosis, can a child be born with something like agenesis or no trachea?

Dr. Karthik Balakrishnan:

Yeah. So, you can have tracheal agenesis. It's a failure of recanalization, we think, of the trachea during embryonic development. This is classified as the Floyd Classification, where in type one, your proximal trachea is absent but you have a distal TE fistula and a distal trachea and bronchi present.

Type two your carina is arising directly from your esophagus and type three you actually have separate main stem bronchi coming off the esophagus. You can imagine that these are pretty life threatening circumstances. The mortality is nearly universal.

There have been a few very bold surgical corrections done in the literature. This is perhaps more common, relatively speaking, with VACTERL. And an important thing here is that if you have tracheal agenesis or atresia, sometimes you can have children present with what's called chaos or congenital high airway obstruction syndrome, which has some characteristic prenatal imaging findings that'll give you a warning.

However, if there is a small TE fistula or even a pinhole opening in the larynx, you may not see those characteristic findings and this can be a very unpleasant surprise at delivery.

Dr. Alyssa Smith:

So, moving on to work up of these patients, if a patient comes into clinic or you're seeing them as a consult, what are some important questions on history that you should be asking either the parents or the child if they're old enough or even the inpatient team that's taking care of them?

Dr. Karthik Balakrishnan:

So, as with any patient with airway concerns, I think it's really important to ask about their prenatal history, their perinatal history. Do they need intubation, supplemental oxygen, things like that.

What is their growth and development and feeding them been like. Do they have symptoms of breathing problems. So cyanosis, retractions, apneas, needing CPR, stridor.

Do they have voice concerns? And in the kids who are old enough to run around and play, you can ask parents to compare them to their age peers to see if their activity seems limited by their breathing. Do they have to stop more often to catch their breath, things like that.

In babies, you can focus more on feeding and say do they have to stop feeding frequently to catch their breath because feeding is a very calorie intensive activity for babies.

Certainly you want to know how old they were when they started to have symptoms. You want to know about recurrent respiratory infections, whether they've been hospitalized for those, whether they've been intubated or had a tracheostomy in the past. And then you want to dig into associated diagnoses.

So, things like for instance Crouzon, Pfeiffer and Goldenhar syndrome can be associated with congenital tracheal stenosis or tracheal sleeve. And actually, one study from Seattle a few years ago said to us that 22% of syndromic craniosynostosis kids can have this tracheal anomaly. So it's a lot...

You can ask about a history of congenital heart disease. An enlarged heart by itself can cause tracheal compression but also a lot of children with congenital heart disease can have other airway anomalies. You want to ask about things like tracheal esophageal fistula, because that can be strongly associated with tracheomalacia.

And then you want to ask about known vascular malformations, and I mean that as a great vessel, not as in vascular anomalies. But things like pulmonary artery sling which can be associated strongly with complete tracheal mix.

Dr. Alyssa Smith:

And then usually our next up in work up is our physical exam. What should we specifically be looking for on physical exam?

Dr. Karthik Balakrishnan:

So first thing is, as with any airway patient, is to do your primary survey. Does this kid look stable? Are their ABCs okay? If they are then you can take a moment and assess them more. So again, you want to do a craniofacial exam, looking for syndromic findings. You want to look for any neurologic deficits.

You want to listen to their heart and lungs to make sure they have good air movement, that they have normal rate and rhythm and a normally located heart for instance. And I generally would recommend doing all these non-airway things first so that your brain doesn't get satisfied if you find an airway problem and miss something else.

Once you've done all that, then you can do you raspatory exam. You can look at the neck, you can feel the trachea, see if it's normally developed and placed. Larynx, same thing. You can listen to the child's voice and cry. And then you can assess them feeding. Potentially even do something like a FEES evaluation while you're there.

Dr. Alyssa Smith:

And then I assume we would be doing a flexible laryngoscopy, just as our initial evaluation, before any other more... maybe invasive airway evaluation. What are we supposed to be assessing on laryngoscopy?

Dr. Karthik Balakrishnan:

Typically, yes. You would do a flexible laryngoscopy as part of your clinical work up. You want to be careful on kids who have congenital heart disease, particularly if it's uncorrected because you can trigger things bradycardic events and vagal events or laryngeal spasms that might be life threatening.

But assuming that you've thought about that, you want to look for, first of all, other airway anomalies. Again it's really easy to go looking at the larynx and miss something higher up. So, you want you check both sides of the nose. Check the choanae, check the pharynx, tongue base and so on.

Then when you check the larynx, you're going to look for the structural larynx vocal fold motion, things like that. And if you can get a view of the subglottis, that's great.

There are some folks who will do awake tracheoscopy even in infants. I have done that on rare occasions. But typically if they're younger than let's say age of five they don't tolerate it terribly well and you have to move so fast with the scope that you don't actually see anything until later when you review the video. But certainly awake tracheoscopy is also an option.

Dr. Alyssa Smith:

And then how about imaging? Is there any role for obtaining x-rays or CT scans?

Dr. Karthik Balakrishnan:

There certainly is. Often these kids will come with an x-ray, it can be useful. You can see obstructive hyperinflation, narrowing in the tracheal air column. But you might not. And if they're crying and their neck is bent or their head is turned or whatever, it may not look particularly clear what is going on.

You have other options. Airway thoracoscopy can be used. It can show dynamic changes. For instance with tracheomalacia. It may be useful in kids who can't tolerate bronchoscopy and so on. But it's less sensitive for mild compression. You can add to its utility by doing bronchography with contrast.

This is done more often for instance in Europe where for very tight tracheostenosis where you can't get an endoscope through, they'll actually drip some contrast on the airway and do fluoroscopy to outline the airway and measure it.

CT scan is commonly used, especially in the US. It's great for comprehensive evaluation. You can do 3D reconstruction. And importantly, if you do with contrast, you can also get detail about the great vessels of the heart, given that we know that many of these tracheal anomalies are associated with heart and vessel abnormalities.

However, it's really hard to get dynamic data in the trachea with that. Videofluoroscopic Swallow Study or it may be useful for concerns of aspiration. But the other thing that's often going to be useful is things like an esophagram where you can look for indentation of the esophagus suggestive of vascular ring.

Dr. Alyssa Smith:

And then I know a lot of these etiologies for tracheostenosis, like the complete tracheal rings and the sleeve that we talked about are associated with syndromes, do you refer all these patients for a genetic evaluation as well?

Dr. Karthik Balakrishnan:

I often do. We're in the process of investigating that. We don't know whether isolated tracheostenosis is associated with any specific genetic anomaly. But certainly if there's suspicion for any sort of syndromic presentation it's absolutely worth doing that.

Dr. Alyssa Smith:

And so usually next ups are going to the operating room for a more comprehensive airway evaluation. And I know we often do flexible and rigid bronchoscopy. Can you speak to the role of each of those methods and the benefits of each?

Dr. Karthik Balakrishnan:

They're complimentary. They certainly don't replace each other. Flexible endoscopy shows you airway dynamics. So for things like malacia and vascular compression they can be very useful. Another nice thing about flexible endoscopy is that you can assess what the airway looks like with different levels of PEEP.

If you use a bronchodapter through an endotracheal tube with the scope, which is very helpful to figure out how bad is the compression or collapsibility.

Flexible bronchoscopy also allows you to look more distally than most rigid scopes will allow you to do. And it lets you do things like bronchoalveolar lavage, if you're worried about post obstructive infections, aspiration and so on.

The advantage of a rigid airway endoscopy is that it allows you to get perhaps more anatomic or structural detail. You get better pictures. You can stent the airway open and push things out of the way if needed and you can also palpate and get some tactile feedback if you're worried about whether a stenosis is firm or soft for instance.

Again, these are complimentary examinations, rather than doing one or the other.

Dr. Alyssa Smith:

And then, can you describe what we should be seeing with a normal trachea. What are normal findings in our exam that we should kind of have in the back of our head for what we're looking for?

Dr. Karthik Balakrishnan:

Anytime you do an airway evaluation you want to really look at the entire airway, right from the tip of the nose down to the bronchi. So you want to look at all those things, but since we're focusing on tracheostenosis today lets assume you've looked at all those.

Normally, you should see horse shoe shaped cartilage rings that take up about four to five out of six parts of the circumference of the airway with the remainder formed by the muscular membranous posterior wall. The rings should be distinct. You should see fine vessels in the mucosa and you should be able to trace the lumen all the way down to the carina and beyond.

Dr. Alyssa Smith:

And then what are some abnormal findings that we might see?

Dr. Karthik Balakrishnan:

Starting from the top, so in kids with congenital tracheostenosis you might see perfectly circular or O-shaped cartilage rings with no membranous trachea. You may see tracheomalacia which you would typically in a child again see a wide cartilage arch with a wide posterior membrane and this posterior membrane will collapse inward with the respiratory cycle.

You may see vascular compression, again half to two-thirds of the way down the trachea on the anterior wall you'll see innominate compression. You usually see a mild degree in most children but it's often not significant. But if there's other kinds of vascular compression you might see compression from different locations including the side or posterior aspect.

You might see a sleeve trachea which looks quite smooth rather than complete rings, but still circular. And then another thing to look for is what's called a bronchus suis or a pig bronchus which is basically a right upper lobe bronchus that takes off of the main trachea rather than the right main stem bronchus.

This in and of itself is not a problem but it's something you'd have to consider if you proceed to do any kind of airway surgery.

Dr. Alyssa Smith:

And then when we're evaluating the stenosis that's present, how do you go about measuring the length or the diameter in evaluating the shape of it?

Dr. Karthik Balakrishnan:

That's a very important question. We really need to be systematic about how we evaluate airway stenoses because the language has to be common across all of us so we know what we're talking about and can communicate with each other.

So, it's important to look at this in terms of severity. And when I say severity I mean radial narrowing. There is no validated system for measuring tracheal stenosis as opposed to subglottic stenosis which typically uses the Myer-Cotton scale.

However, the general consensus is that the Myer-Cotton scale is also useful for tracheostenosis and that's what most of us are using now until something better comes along.

You also want to look at the length of the stenosis and you can measure that in centimeters often by marking your bronchoscope as you slide it in along the length of the stenosis.

You want to look at the shape, is a circular, is it tortuous corkscrew shaped. The character, is it firm or soft. And does it appear to be cartilaginous or scarred, for instance.

And then you want to look at the location. Does it involve the carina, does it involve the larynx, is it somewhere between. So those would be kind of the main things.

There are a lot different ways to do this. People are using imaging software and all sorts of things. But again, a lot of us just keep it very basic here.

Dr. Alyssa Smith:

And so you mentioned the Myer-Cotton grading system and I know that we talked about this the subglottic stenosis episode of the podcast, but can you go over briefly what that grading system is again?

Dr. Karthik Balakrishnan:

Absolutely, so again keep in mind this is developed for the subglottis but often applied to the trachea. Essentially it says that if you have a stenosis of 0-50% that's grade one, 51-70% is grade two, 71-99% grade three and grade four is a complete occlusion.

Dr. Alyssa Smith:

So after evaluating the trachea I will often take a look down at the bronchi as well. How far do you look and is there anything in particular that we're looking for in the bronchi?

Dr. Karthik Balakrishnan:

It is really important to see the airway as one unified structure so you're absolutely right that we should look at the bronchi as well.

When you're doing this it's really important not to try to force your bronchoscope or anything else through a stenotic segment because you could actually cause mucosal trauma with more edema and worsened obstruction.

Particularly in congenital tracheostenosis it sort of has a funnel shape and the distal segment is most stenotic so if you try to shove something through there you can create a critical or potentially unsalvageable airway.

That said, if you can get past that area to look at the carina and bronchi, it is a wise thing to do. You'll sometimes see bronchiostenosis in the setting of tracheostenosis.

Tracheomalacia may be associated with bronchomalacia in up to 30% of kids. And you also want to look for things like post obstructive infections, inflammation and so on.

Not to mention, I should say, if you have vascular compression of the airway, sometimes you can have that in more than one location as well.

Dr. Alyssa Smith:

Right. And then moving on to our various treatment options. For our patients with mild symptoms, can these patients be managed conservatively?

Dr. Karthik Balakrishnan:

Absolutely. It kind of depends on the cause of the stenosis or narrowing. If you have mild tracheomalacia, sometimes those kids need nothing at all.

A little bit more than that they might need antibiotics for prolonged or recurrent respiratory infections. They might need inhaled steroids, sometimes things like bethanechol to cause essentially tension of the posterior membranous trachea to keep it open.

If it's more of a stenosis than a malacia, the very mild kids will often outgrow this problem given time. But again they might be a little more prone to worse symptoms with respiratory infections. They might need humidification, intermittent steroids, chest physiotherapy for both malacia and stenosis.

Dr. Alyssa Smith:

So, thinking about the surgical options that are available, can we talk about different options for each of the different causes of tracheostenosis?

Dr. Karthik Balakrishnan:

Sure. So let's say that you have a purely endoluminal stenosis, that's either a congenital... something like a tracheal web or an acquired stenosis. If it's a pretty short segment, then it may be amenable to balloon dilation.

Typically, that's going to be ideal for sort of a thin web like mature scar. Sometimes you can combine that with things like radial incision, steroid injection and balloon dilation to get more effect.

While there's not clear data to guide us, a lot of people would try that two, three, four times before they say it's not working. If it is a fairly short segment and either to amenable or not responsive to dilation you can also consider resection.

That length that you can resect is pretty limited depending on the patient, so in most children I would say it's 25-30% of the tracheal length. So, in a premature baby or a new born that's about a centimeter. In an infant it's about a centimeter and a half. So it's not a very long length.

So those are options, and if you have a longer segment stenosis then you might try a variety of things. Slide tracheoplasty is what I would typically use if it's a significant stenosis that needs intervention. But other people have huge patch tracheoplasty, they've used grafts with cartilage and even in extreme circumstances replacing the trachea with things like allografts.

Dr. Alyssa Smith:

So let's talk a little bit about slide tracheoplasty. What are some of the benefits of this approach compared to just the resection and reanastomosis?

Dr. Karthik Balakrishnan:

Slide tracheoplasty is great because it's very adaptable. It has some significant advantages in that unlike graft tracheoplasty you are using native tracheal tissue unlike a simple resection and reanastomosis.

You are distributing the tension over a very long suture line and again unlike resection and reanastomosis, instead of creating a circular anastomosis that might scare back down, you're creating an oval anastomosis. That's less likely to do so.

So these are all really important things, and really though I think the biggest advantage of this is that you're not limited by length. You can really slide a child from the cricoid well past the carina, unlike a resection.

Dr. Alyssa Smith:

And I know this may be difficult without any pictures or drawings or anything but can you try to describe exactly how this is done?

Dr. Karthik Balakrishnan:

Sure. So it is certainly a complex procedure but the basic geometry is if you imagine a paper towel roll as the trachea and let's say the whole length is stenotic.

So you're going to transect the paper towel roll or trachea 50% of the way along its length. So at the mid point of the stenosis. Then you're going to filet open the anterior surface of one half and the posterior surface of the other half and slide them over each other so that you end up with a tube that's twice as wide and half as long.

Dr. Alyssa Smith:

And I can imagine that your approach might be different depending on exactly where the stenosis is in the trachea.

Dr. Karthik Balakrishnan:

Correct. So, you can access the upper half to two thirds of the trachea through the neck, and if you do that then you would typically... once you divide the trachea intubate through the neck wound in to the distal limb intermittently while you're fixing things.

If you're going out anymore distal than that, to the distal trachea, corina and main stem bronchi then typically a different approach is required. The most common approach nowadays is to do a median sternotomy and this repair either on ECMO or bypass.

There are people, including Dr. Grillo who is a real leading thoracic surgeon who would do this through a thoracotomy without bypass, but most people nowadays will do it via sternotomy with some sort of cardio pulmonary support.

Dr. Alyssa Smith:

And then if a resident is able to be involved in a case like this, do you have any key technical pearls or pieces of wisdom that you can pass on?

Dr. Karthik Balakrishnan:

Sure. So, a couple of things. The first is that typically you're going to do the anastomosis with a running double arm suture so that it's a single stich doing the entire anastomosis so be gentle with the stich and don't blunt your needle, don't grab the tip of the needle.

Another key thing is eversion of the edges of the cartilages as you're sowing them together. You can imagine again your paper towel roll that you've split the front of one half and the back of the other half and you're sowing them together to make a bigger tube.

You can picture that they would... each half would want to curl back in on itself to resume its original circular shape and that's called a figure eight deformity. Cartilage does the same thing. And the best way to avoid that is as you're tightening each loop of your stich you want your partner across the table to evert the edges of the cartilage to minimize that risk.

Keep in mind too that if you're doing this via thoracic approach you're working through a pretty small window. So you're kind of working on the distal trachea through a square that's made by the superior vena cava on the right, the aorta on the left, the pulmonary artery and the innominate vessels superiorly. So you have a pretty small box to work through so knowing your regional anatomy is critical here.

Dr. Alyssa Smith:

And then for children that have concurrent cardiac anomalies, are these usually repaired simultaneously or is it a staged repair?

Dr. Karthik Balakrishnan:

It depends on the anomaly. Typically, they're repaired at the same time because you've already got the sternotomy, you've already got the access and the child is going to go on pump. So it's often best to do that at the same time if you can.

Dr. Alyssa Smith:

And then are there any kids that wouldn't be a good candidate for a slide tracheoplasty either based on their underlying medical conditions or the characterization of the stenosis itself?

Dr. Karthik Balakrishnan:

There are. They are a few. Slide tracheoplasty is adaptable enough you can apply it to almost anyone but the cases where it would not be useful is if you have no usable trachea lumen, so let's say you had a complete tracheal agenesis, you have nothing to slide it with.

Similarly, if you have absent trachea rings, that segment cannot really be slid so it has to be, at least partially, resected. You can use slide tracheoplasty for grade four stenosis with no lumen but again that's not ideal. So if you can resect the worst part of the stenosis and slide the rest, that's better.

There are some kids who are just too sick to go through this, to be honest. If you get to the point where you're considering it, you may not have a lot of choice because they have no salvageable airway otherwise.

Dr. Alyssa Smith:

And then looking at tracheomalacia, I know we had talked about some conservative measures. Are there any surgical interventions that you can do for these kids that have more severe symptoms?

Dr. Karthik Balakrishnan:

Yeah. The mild kids you can manage with things like CPAP or medications like we discussed. More severe, the traditional treatment has been tracheostomy and potentially administration of positive pressure through that.

But that's not always an easy thing for families to adapt to. Aortopexy is an option. The idea there is that you basically approximate the aorta to the posterior sternum and the soft tissue attachments between the aorta and the trachea therefor pull anterior wall of the trachea forward widening its lumen and again as we discussed earlier narrower airways tend to collapse more conversely wider airways collapse less and so you've reduced some of that collapsibility with respiration.

Stenting is an option and we can talk a little bit about different kinds of stents and indications. It's not an ideal option for a variety of reasons. And then in really bad cases if it's a segmental portion... A segment of the trachea you can potentially resect it or slide it.

Or in some rare cases we've even done external bracing with cartilage grafts or plates.

Dr. Alyssa Smith:

So you mentioned that there's different types of stents that can be used. Can you elaborate on that a little bit and talk about the different types?

Dr. Karthik Balakrishnan:

Yeah. So there are a huge variety of stents and it's increasing all the time but if we break them into general buckets, the first is what you could call balloon expandable stent that are typically metal mesh.

One common example of this would be a [polmile 00:30:47] stent. The nice thing about these is that you can adjust their diameter, you can conform them to tortuous airways, you can flair them so that the ends dig into the mucosa so that they don't slide.

And the epithelium can actually grow through the mesh work so once things heal up you don't loose ciliary function quite as much. However, these can provoke a lot of granulation because they get

incorporated into the tracheal wall after six or eight weeks, removing them can be very problematic and damaging to the mucosal area, even the wall of the airway. And they can erode into adjacent structures.

Another option is a covered mesh stent, so one common example would be an eye cast stent. And the idea there is again you have this metallic mesh with all the advantages in terms of being able to flair it and shape it in and adjust its diameter with a balloon but it has a coating over it so that you can't get tissue ingrowth so it's a lot easier to remove.

The draw back there is that you can get biofilms on the internal aspect of the stent which can lead to plugging and mucus buildup. Sort of with the same types of problems as that coated stent or silicone stents so the Dumon, the hood stent, cut T tubes, things like that. They're pretty easy to place. Depending on their shape they can either hold themselves in place or you have to secure them with a suture.

They're pretty non reactive and so you don't have a lot of granulation formation and so on often. But there's no mucociliar clearance or ciliary function for that stretch of the airway. They don't conform to the airway well.

They can have biofilm and plug and they can migrate because they're smooth and slippery. Which is why sometimes you need to suture them. A new area that's emerging is what area called bioabsorbable stents. So things like the ELLA stent.

These were developed for esophageal use. They're typically self expanding and made of [PDS 00:32:34] for the most part. They can be custom ordered for use in the airway and they're used a fair bit in Europe for that purpose.

The nice thing about them is they last six to eight weeks and dissolve away. And if you need to place a new one you just place it but you don't have the problems of having to remove the stent and damage the airway as you do that.

Dr. Alyssa Smith:

And then finally, quickly discussing vascular rings, what do we do about that? Is there any surgical treatment for vascular rings?

Dr. Karthik Balakrishnan:

Yeah, vascular rings are often managed surgically. Typically, in collaboration with our cardiovascular surgery colleagues. Depending on the type of ring you essentially divide the ring and create space for the airway. So if you have a double aortic arch, usually the right side is dominant, so you get rid of that left side.

If you have a pulmonary artery sling that's causing significant problems then you can either divide the sling and re-implant the pulmonary artery or if you also have to repair the trachea then sometimes you can divide the trachea and transpose it anterior to the pulmonary artery before you do you slide and anastomosis.

Dr. Alyssa Smith:

So focusing specifically on open airway surgery, what is the typical regiment for a patient postoperatively?

Dr. Karthik Balakrishnan:

The most critical thing is to keep in mind what are the things that can go wrong. So one thing that can go wrong is you can over pressurize the repair with your mechanical ventilation and blow up in the anastomosis and cause a partial or complete dehiscent. So we want these kids spontaneously ventilating or negative pressure ventilating as soon as possible.

Another thing that can happen is you can get granulation from the ET tube so again we try to get these kids extubated as soon as we can, hopefully within a day or two if they do well.

Third possibility is, you've got this suture line in the airway that can accumulate clot or mucus because you don't have ciliary function right at that point. And that can be prevented with high humidity and if the child is intubated then frequent drips of selene down the ET tube with suctioning as well.

Those are some of the key things that can happen. We also like to do a follow up endoscopy, usually a week after surgery to look for things like that figure eight deformity and granulation tissue and so on. And then obviously we have to be careful about starting FEES afterwards until we're sure that the recurrent nerves are working since those run right next to the trachea and might be injured.

Dr. Alyssa Smith:

And then what does long term care and follow up look like for patients that have undergone open airway reconstruction?

Dr. Karthik Balakrishnan:

So these kids do need long term follow up. They need routine airway endoscopy and the idea is that we want to make sure that anastomosis continues to grow with them, that their airway continues to grow with them.

So typically, I will bring kids back a week after surgery, a couple weeks after surgery, a month after surgery, three months after surgery, six months, a year and then do it every six months for a couple of years and then space it out after that.

That's just my routine. A lot of people do it differently but regardless I think everyone who does this kind of operation is going to be following these kids over the long term.

Dr. Alyssa Smith:

And then when you're talking to parents, preoperatively and you're counseling them on the risks of surgery, where you talked about suture line breakdown and granulation tissue formation, figure eight deformity, what about the long term rate of restenosis? What do you kind of counsel parents about when you're talking to them about this?

Dr. Karthik Balakrishnan:

There's not a huge amount of data on this but in general if you do a side tracheoplasty successfully, let's use that as an example, then the rate of restenosis is actually quite low.

I think the most common reasons that you would need to go back and re-operate or re-intervene is either during the initial postoperative course if things don't go perfectly or if some pathology was missed. So for example if you do a slide tracheoplasty or resection but you miss some area of the stenosis that's still there.

Dr. Alyssa Smith:

And then as far as the neutral history of tracheal stenosis goes, what would likely happen to these patients should no intervention be performed?

Dr. Karthik Balakrishnan:

That's a really important question and something that's really important to counsel parents about. So let's start with tracheomalacia. Tracheomalacia typically gets better with age.

It sometimes worsens in toddler-hood because the child starts being more active and puts more demand on their airway and is breathing harder.

As typical congenital tracheomalacia will improve by age two to three years, sometimes it can take longer. It's important to warn parents that until things improve, as the child grows, the child may have frequent hospitalizations, may have prolonged symptoms when they have respiratory infections, may have more difficulty with activity and so on.

If the child is syndromic then perhaps they are more likely to have prolonged symptoms into an older age with the tracheomalacia.

If they have more a intraluminal or cartilaginous stenosis of the airway then the symptoms you might expect to get worse with age as they put more demand on their airway and they essentially outgrow their airway lumen.

That said, even kids with complete tracheal rings, the literature suggests that maybe 10-15% of them will never need intervention. But you have to follow them over time because even if they have a growth spurt in their teen years they might start to develop symptoms that... We've had patients before who are college athletes who had undiagnosed complete rings that became symptomatic when they started doing really competitive sports.

Of course if there is a very severe stenosis then the risk to the child in terms of morbidity and mortality is much higher and so then you would counsel more strongly for intervention.

Dr. Alyssa Smith:

So, before we move on to our summary, can you comment on any potential future directions of the field of tracheal open airway surgery, any new advances or new techniques that are being developed?

Dr. Karthik Balakrishnan:

Sure, so there's some pretty cool stuff going on all over the place. One hot topic of course is tracheal replacement. There's a group of superb surgeons in Paris who, a couple of years ago, published in the New England Journal, where they essentially rebuilt a child trachea out of free flaps and cartilage grafts. That's pretty amazing.

Another option of course is tissue engineering and there are people who are working on developing scaffolds and cell seeding to do that.

Tracheal transplantation is something that's in the works as well. And of course robotics is a hot topic, though currently nobody is doing robotic tracheal surgery. We are working on developing that technique right now.

Dr. Alyssa Smith:

Right, so in summary. Patient presentation is dependent on the degree of stenosis. The common presenting symptoms include stridor, wheezing, cough, recurrent respiratory infections and varying degrees of respiratory distress.

Common causes include congenital tracheal rings or tracheal sleeve. External compression from vasculature or other mass. Dynamic collapse due to tracheomalacia, an acquired stenosis from previous intubation or tracheostomy.

Bronchoscopy is the gold standard for airway evaluation in diagnosis. Intubation and manipulation of the stenotic segment should be avoided when possible to prevent edema and additional stenosis.

And often in congenital tracheostenosis, the stenosis is a funneled shape. So the more distal segment is going to be the most stenotic.

There's a high prevalence of associated anomalies which should be kept in mind during history gathering in both bedside and endoscopic evaluation. In patients with mild to moderate symptoms can sometimes be managed conservatively.

All patients with severe stenosis will likely require intervention. Short segments of stenosis may be able to be treated endoscopically but longer and more severe segments will likely require open airway surgery.

Dr. Balakrishnan thank you again for joining us, is there anything else you would like to add?

Dr. Karthik Balakrishnan:

Thank you Alyssa, this was great.

The only thing I'd like to add is just as the listeners can tell, this is a very complex field and these are complex children. I think that it's always worth considering that these might be best managed through a multidisciplinary program like an [aero00:00:40:36] digestive program or a complex airway reconstruction program rather than doping it as a one off.

Dr. Alyssa Smith:

Awesome, again thank you so much.

Dr. Karthik Balakrishnan:

All right, thank you.

Dr. Alyssa Smith:

I'll now move on to the question portion of this podcast. As a reminder, I'll ask a question, pause for a few seconds and then give the answer.

So the first question is what causes the dynamic collapse seen in tracheomalacia?

So, during exhalation, extraluminal pressure exceeds the intraluminal airway pressure and in tracheomalacia the weakened cartilage is unable to withstand this pressure differential which leads to collapse of the airway.

The second question is what are some common symptoms in patients with tracheostenosis? Some common presenting symptoms are stridor, wheezing, cough, and varying degrees of respiratory distress.

Our third question is what syndromes are associated with tracheal sleeve and what vascular anomaly is most commonly associated with complete tracheal rings?

Tracheal sleeve is associated with Crouzon, Pfeiffer and Goldenhar syndromes. And then complete tracheal rings are associated with pulmonary artery sling.

Thanks so much for listening and we'll see you next time.