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 on to 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 otolaryngologist Dr. Paul Willging. In this episode we'll be discussing velopharyngeal dysfunction, specifically focusing no velopharyngeal insufficiency. Thanks for being here, Dr. Willging.

Dr. J. Paul Willging:

My pleasure, thanks for having me.

Dr. Alyssa Smith:

To start, can you just discuss what velopharyngeal dysfunction is?

Dr. J. Paul Willging:

Velopharyngeal dysfunction can kind of be divided into two different pieces. It's a problem with how the soft palate opposes the posterior pharyngeal wall. We need good function on that to have normal residence and normal speech production. So velopharyngeal dysfunction presents itself as abnormal speech, abnormal resonance, and a difficulty with understanding what someone says.

Dr. Alyssa Smith:

When thinking about presentation, how does a patient with velopharyngeal insufficiency typically present?

Dr. J. Paul Willging:

The families will often say that the child talks through their nose a lot. They will sometimes complain about, the child is always mumbling and they can't understand what the child is saying. When you listen to the kids you hear some abnormal resonance. Families will often mistake hypernasality and hyponasality. So if the child has a very large adenoid pad, they're very stopped up, they can sound hyponasal, and many families will feel that is abnormal resonance, which it is. But it'll be confused for hypernasality. So we're looking for abnormal resonance, we're looking for hypernasality, too much resonance and sound coming through the nose. It's not a stopped-up sound, it's an overproduction of the nasal sounds.

Dr. Alyssa Smith:

Is there a typical age at which these children may present?

Dr. J. Paul Willging:

The age of three is frequently when you can accurately make the diagnosis for VPI, velopharyngeal insufficiency. You need the child to have connected speech so you can hear how they sound when they're speaking in sentences. A child may have significant VPI but be able to make individual words, but when they put them in combination things will break down very rapidly. Most children, by the age of

three to three and a half, will have enough connected speech that you can do a proper velopharyngeal sufficiency evaluation.

Dr. Alyssa Smith:

Before we think about the abnormal development of speech, or VPI, can we first discuss normal speech production and what structures are involved in articulation?

Dr. J. Paul Willging:

We tend to break it down into an energy source, so you need your larynx to produce the sound. You then have some resonating chambers. You have the pharynx, you've got the oral cavity, and you've got the nasopharynx, as being those resonating chambers. Then you have your articulators, which would be the lips, the tongue. Tongue position and tongue shape can have an effect on that as well. And the velum needs to be in its proper position to properly produce these different sounds as well.

Dr. Alyssa Smith:

You mentioned the positioning of the velum is important for proper production of sound, but how exactly is the velopharyngeal port controlled?

Dr. J. Paul Willging:

During speech the soft palate elevates and pushes back to make contact with the posterior pharyngeal wall. The side walls come in and they form a good seal, effectively separating the nasopharynx from the oropharyngeal down below. The soft palate needs to be in that upward and sealed position for all sounds in the English language. The only exceptions are the M, N, and N-G sounds, which are nasals. To properly produce those, the soft palate needs to be down.

Dr. Alyssa Smith:

With that in mind, what causes a patient to develop VPI?

Dr. J. Paul Willging:

We can have some structural problems. A child can have an overt cleft to the secondary palate, so the child's unable to get things to close because of that large gap in the central part of the soft palate. You can have some other structural problems, where there's a submucous cleft palate. So the musculature has done exactly the same as on an overt cleft. The levator muscles are coursing anteriorly to attach to the posterior border of the hard palate, and the levators can't elevate and posteriorly displace the soft palate. So the structural problems of an overt cleft or a submucous cleft palate can cause it. If patients have had some type of tumor extirpation where they're missing part of the soft palate, that can also cause it.

There can be some neurologic problems, where there's problems with the brain stem, where things just are not functioning well. Some of the muscular dystrophy patients can have difficulty with tiring out, the myasthenia patients will often have problems later in the day because again, they tend to tire out. So we have structural problems and neurologic problems.

I guess there's an in between zone, where the structure looks good, neurologically everything is okay, but the child from a functional standpoint just cannot get things to close. Some of the syndromes, like velocardiofacial syndrome, the anatomy is normal, the neural structures and function all look

normal, yet the kids just have this generalized hypotonia, or hypopharyngeal hypotonia, and they just cannot get the soft palate to seal up against the back wall of the throat.

Dr. Alyssa Smith:

I think as residents, we all learn about VPI as an important complication of adenoidectomy. Can you touch on that a little bit?

Dr. J. Paul Willging:

I think looking through the literature, the overall incidence of VPI following adenoidectomy is around one in 2,000. Those patients who develop VPI postoperatively, if you go back and really look at those kids, and kind of delve into their past history, you'll usually find that the kids at risk for VPI following adenoidectomy are those who have a family history of clefting. Because the potential for a submucous cleft in that particular patient is higher. The kids may have a bifid uvula that was not identified. They may have a notch in the posterior border of the hard palate, which could identify a submucous cleft.

Looking at the functional side, the kids who at a young age had significant feeding difficulties as an infant may have some trouble if you take their adenoids out. Children with some generalized hypotonia can have some increased difficulty if you do a complete adenoidectomy. I feel you can still do an adenoidectomy in children with a history of a cleft palate, with a history of a submucous cleft palate, with a history of feeding problems. But in those kids we will frequently leave a rim of adenoid tissue inferiorly, so we'll kind of do a superior half-adenoidectomy. We remove the tissue that may be blocking the posterior choana, interfering with eustachian tube function, but we still leave the bulk of the tissue inferiorly so the soft palate can hit up against the posterior wall as it was in that preoperative condition.

Dr. Alyssa Smith:

Moving on to workup for these patients, what are some important questions to ask when gathering a history?

Dr. J. Paul Willging:

Evaluating a birth history, was there some type of anoxic problem going on with delivery. How was the child feeding at an early age. Then, when the family has noticed some abnormal speech development. So the history may direct you toward a neurologic problem if there were some significant birth questions going on. You want to know if the child has had any previous surgery, like a tonsillectomy and adenoidectomy. Certainly if they've had a cleft repair, you would want to know about that, because the incidence of VPI following cleft repair is approximately 20%. So that's going to tip you into the structural problems as well. So the history's just going to let you early on determine, does this child have some neurologic things going on, are there any syndromes that you need to be aware of, and has there been any surgical intervention done prior to the development of VPI.

Dr. Alyssa Smith:

Next, can you walk us through your approach to the physical exam for these patients?

Dr. J. Paul Willging:

In my particular clinic, we do it in a multidisciplinary center. So we have speech pathology, genetics, and ENT. In my clinic we are looking at the non-cleft VPI patients. So when they come into our clinic, I'm going to a examination of the head and neck. We're looking at nasal structures, we're looking in the oral

cavity, looking for a bifid uvula, signs of a submucous cleft. We're getting a history of whether or not the child has met their developmental milestones on time. If there's developmental delays it's kind of a trigger that there might be some developmental problems going on, some syndrome involved with this. So the physical examination's not very involved. Most of our information's going to come when we put the flexible scope in.

We always get a speech sample on the child before we start to agitate them, before we put the telescope in. So as an otolaryngologist, I can listen to the child, I can tell if they have hypernasality. But we have our speech pathologist do that, who can also be aware of what normal developmental substitutions would be. They can look at the generalized oromotor skills that the child has. They can also do some therapy sessions right there, and see if they can get the child to improve any hypernasality or nasal air emission that they hear. Is this a child who's having some mis-learning as the source of their VPI, or is this a persistent problem that therapy is not going to be effective for? We generally have our genetics people see the patients after we've done the flexible scope, and they do a much more detailed birth history, and kind of a complexity examination looking for any potential underlying syndrome.

Because my clinic is not looking at the overt cleft lip and palate kid, we have a very high percentage of patients coming to my clinic being identified with an underlying syndrome, velocardiofacial syndrome being the most common. So my patient population is different than the generalized cleft group. That's why we have genetics involved in my clinic.

Dr. Alyssa Smith:

You mentioned performing flexible endoscopy for these patients. Can you discuss what specifically you're looking for in evaluating?

Dr. J. Paul Willging:

A lot of centers feel like you really can't do a good assessment on young children without sedating them. If you sedate a child then you're trying to look at velopharyngeal function. If it is abnormal, is it because you've sedated the child, or they have a functional problem? So we do not sedate any of these kids, we just do it in our regular ENT clinic. I will put some topical aesthetics in the nose. We have a little mist bottle. I put a one-to-one mixture of Afrin and 2% pontocaine into this, and then we just atomize some material up into the nasal passages. You want to make sure that you're spraying vertically so we're not going to anesthetize the soft palate, so spraying vertically we're getting the inferior turbinates and the septum in the middle meatus area. And the cooperation level goes way up if you can topically anesthetize these kids. I'll then put some 2% viscous lidocaine on the endoscope, just so anything that the scope physically touches gets anesthetized a little bit more.

So we pass the telescope through the middle meatus, because we want to be looking down onto the velopharyngeal sphincter area. If we go along the floor of the nose, even though the area's sometimes easier to pass through your scope, you're going to be looking at the back wall, you're going to be looking tangent to the velopharyngeal closure area. So when we go high through the middle meatus and look down, we're looking at palatal structure. If there's a submucous cleft palate we can see a muscular bulge coming anteriorly with that zona pellucida, that just nasal mucosa and oral mucosa in the midline. So you can see this trough in a child who has a submucous cleft.

Our speech pathologists are then working with us to get the kids to communicate with us. They have a standardized set of phrases to make the velopharyngeal sphincter get stressed a little bit. We're looking at closure, we're identifying where the kids are having trouble closing, in other words where's the hole. Is it a central gap? Is it across the entire velopharyngeal sphincter area? Is it one lateral corner? Is it bilateral corners? So we're just trying to identify where the leakage is coming from, and how

consistent it is, and how big it is. We can't really quantify the size of the hole, but you can certainly describe its location, and is it a small transient thing which intermittently is leaking, is it a persistent opening, is it a real large opening. Because that's going to have an impact on how you're going to surgically correct these.

Before I take the telescope out, I always go down and look in the hypopharynx. I want to know how the vocal folds are moving. I want to look at the vocal folds to see if there are any vocal fold nodules. Many of these kids with hypernasality will compensate for some of their inability to close the velopharyngeal sphincter by making glottal stops. That's very hard on the larynx and they tend to develop vocal fold nodules. You correct the VPI, the nodules tend to go away. Because of the high incidence of syndromic patients in my clinic, and velocardiofacial syndrome is the most common, those kids can also have medialized carotid arteries. So I'm always looking in the pharynx to see if I can see these medialized carotids pulsating on the back wall, and I always note on our surgical request form if I see the vessels, and kind of which side they are, just so you can be aware when you're going in there.

Dr. Alyssa Smith:

You mentioned that there can be different patterns of closure when you're taking a look at the velopharyngeal port. How do you describe these different patterns, and do they vary in frequency?

Dr. J. Paul Willging:

Probably the most common is a coronal closure pattern, where the soft palate is coming straight back and hitting the posterior pharyngeal wall. That's probably 60% to 70% of the kids, will have that type of pattern. There's a circular pattern, which is probably the next most common, where the soft palate is coming back in the coronal plane, but the posterior wall is coming forward a little bit, and the side walls are coming in a little bit. So you kind of have contributions from all different directions closing the velopharyngeal port. There's also the sagittal closure pattern, where the side walls make up almost all of the closure activity which is going on. The palate elevates, but doesn't really translate posterior at all, and it's just the side walls coming in.

Dr. Alyssa Smith:

Are there any other methods of testing that can be used, either that you use in your clinic, or historically that were used?

Dr. J. Paul Willging:

I guess prior to endoscopes being available, they used to use some mirrors, just like checking for choanal atresia. You take a cool mirror and put it in front of the child's nostril and have him repeat some sentences, and if you had a lot of air escaping from the nose your mirror would fog up. There should be some nasal escape in many connected speech sentences, so it's a very inaccurate type of a test, it's really not used much at all any more. There are some radiographic tests that you can get. You can get some still films, some cephalometric tests. It lets you measure the distances from different structures, but it doesn't tell you the functional aspect of how the kids can close the velopharyngeal sphincter.

There's the videofluoroscopic speech study, where barium is instilled into the floor of the nose, you have the kid sniff it back, and it coats the soft palate, the posterior pharyngeal wall, the lateral walls, and then you get a lateral fluoroscopy view when the child is repeating several standardized sentences. You can also turn the child in the AP direction to see lateral wall motion, and then you can also do what's called a base view, where it's coming from the submental area up through the velopharyngeal

sphincter, to look at the proper plane of closure. It's very difficult to get the kids to cooperate with the videofluoroscopic speech studies. Barium burns in the nose.

We used to do both the fluoroscopy studies and our nasal endoscopy back in the early 1990s, and trying to compare what we saw in the fluoroscopy studies, what we saw in the nasal endoscopy exam, and seeing if we were missing anything on either of the exams. We could see everything that we needed on nasal endoscopy, so we don't do the videofluoroscopic studies any longer. The benefit of the X-ray study, the fluoroscopy studies, are that you can measure the size of the gap. Some of the problems with the fluoroscopy studies, it's very easy to miss small gaps. Because you're looking at the sum of the view going from left to right on the lateral, you can have a unilateral gap and you will not see it on the lateral exam. If you do the base view, you will be able to pick that up. Sometimes you can pick it up on the AP view. But some of these gaps are small, and if you're not really good on the technical side it's easy to miss these. So we've kind of limited our examinations now to just the nasal endoscopy.

We'll still get some fluoroscopy studies if we have children with cul de sac resonance. That's kind of that hot potato voice that you see with a peritonsillar abscess on steroids. They're obstructed in the hypopharynx, and you can't see that looking with a nasal endoscope. So in those kids with cul de sac resonance, which your speech pathologist will be able to tell you that's what they're hearing, you'll still get the fluoroscopy studies on them.

Dr. Alyssa Smith:

Can you touch on what nasometry is, and what information it provides?

Dr. J. Paul Willging:

It's nice to have objective measures preoperatively so you can compare it to your post-op results. Nasal endoscopy is a device that [inaudible 00:21:24] Metrics had developed. It is essentially a plate that goes on the upper lip, and there's a microphone above the plate which is measuring, essentially, sound coming through the nasal passages. There's a second microphone beneath the plate, which is measuring sound energy coming through the mouth. You have the kids repeat some standardized sentences, and then you can take this ratio of sound energy coming through the nose to sound energy coming through the mouth, and you end up with what's called a nasalance score.

You really have to know what you're listening to in order to interpret what the nasalance score is. If you have a child with a very tiny gap, they're going to have a lot of high-pressure air coming through a small area, and this jet of air, essentially, is going to cause a lot of turbulence and it makes a lot of noise. And you're going to get very high nasalance scores, but it's a very small gap. You can have a child who has horrible velar movement, so it's an extremely large velopharyngeal gap. They essentially have no separation between the oral passage and the nasal passages. There's almost no sound coming through the nose, it's just the resonance coming through. So those kids will actually have a huge gap, but a much lower nasalance score. So it still tells you there's abnormal resonance, but the score does not really correlate with how big the gap is. So you can have much worse nasalance scores postoperatively, even though you're really close to getting them back to normal resonance. So again, you have to interpret the nasalance scores based on what you did and where the gap is, so you just have to be careful with it. But it does give you objective information.

Dr. Alyssa Smith:

You've mentioned quite a few times how many other services are involved with your VPI clinic. Can you just speak to the benefit of having a multidisciplinary VPI clinic?

Dr. J. Paul Willging:

As a surgeon, we are looking at these patients with the bias of, you have velopharyngeal insufficiency that I hear, and I have an operation that can fix you. I think it gives more credence to your argument if you have an independent assessment of speech quality. A speech pathologist is going to tell you they cannot fix certain types of problems. So we actually get many more referrals for speech surgery coming from our speech pathologists. They are assessing the child, they're telling the family that their child has intelligibility problems. The families are usually relieved to hear that it's not just them, their concerns are being corroborate. And as a surgeon we are then going to go in to identify what the problem is and how we can fix it. So it just kind of gives us an additional pair of eyes looking at the child to say we do have a problem.

There are some kids that, as an otolaryngologist, I hear significant hypernasality. There's a lot of nasal air emission. Every time the child makes an S sound, horrible noise comes through the nose. The speech pathologists will do their nasometry, and they have horrible scores, but it can sometimes be mislearning. The way the child learned to make the S sound is just improper. And with some therapy techniques that very quickly teach the kid how to make that sound correctly, they can take care of it.

So my speech therapists like to get me all excited because there's a child with horrible VPI, they're making the child do all the S sounds, and I'm watching this bubbling come across the entire posterior velopharyngeal sphincter area, and I'm smiling to myself thinking we're going to get the flap in the resonance, and fellas get to do some fun operations. Then they'll start some therapy sessions with the telescope in, they'll tell the child to look and make this sound a different way, and all of a sudden the nasal emission goes away. The families can also look at that nasal endoscopy exam, they can see the air escaping, they can hear the noise, then they see how the therapy works on closing that gap, and the sound starts to normalize.

So I think you just have more credibility in a multidisciplinary team when you recommend surgical intervention that speech therapy is not going to take care of some of these. On the other hand, we also have genetics who can identify some underlying syndromes, and we can anticipate problems that these children are going to have over time. The velocardiofacial kids, for example, have that generalized hypotonia. They will tell me through testing that this child has velocardiofacial syndrome, and I know that I have to be more aggressive on my surgical intervention on that kid. When I inset the flap I have to make the lateral port smaller than I would ordinarily do for an otherwise normal child.

We also know that these kids are likely to have some developmental delays, so our geneticist will get them involved with early intervention, they will get special IEPs for the kids to follow at schools. So we're not waiting for the kids to decline before we get intervention, we're being very early in our interventions for these kids. But that's because we have the multidisciplinary team, and everybody's kind of looking at the kid through a different lens.

Dr. Alyssa Smith:

Next let's talk about treatment. In general, what is the treatment approach for these patients?

Dr. J. Paul Willging:

I think you have to define what the problem is. So are we looking at a misarticulation, a mislearning type of a problem? If it is, speech therapy is what's going to fix these kids, and you do not need surgical intervention for them.

There's going to be a group of kids who have a structural problem, or a functional problem that the therapists are not going to be able to overcome. Those kids, you are looking at either surgical intervention, or you potentially could go down the prosthetic route.

There are two different types of prosthetics that can improve velopharyngeal insufficiency. If you have a neurologic problem where there's paralysis of the palate, so the child had a brain stem lesion, they have a unilateral paralysis, the soft palate on one side is not functioning, or bilaterally the palate's not functioning, you could do what's called a palatal lift. So it kind of looks like a retainer with a bar coming off the back edge of it to push the palate up and back. It's a static device, it works well in the flaccid soft palate. If there's a lot of motion, if the kids can drop the palate, they generally tend to push the prosthetic off the tooth anchors. So it works well in the paralysis kids, does not work well on the neurologically functioning palate.

There are obturators, which kind of look the same as the retainer. It has a bar coming off the back, and then it has a speech bulb that goes up into the velopharyngeal port area. And that can fill in that gap that the children are unable to close.

It takes a prosthodontist, and as otolaryngologists, we use prosthodontists for a lot of different things, for our head and neck cancer reconstructions. So those individuals will frequently be able to make some of these prosthetic devices for us. The devices are relatively expensive. The devices have to be worn all the time to be effective. The devices can frequently cause a fair amount of discomfort for the kids, they can get some choking and gagging because this big speech bulb is going up through the posterior pharynx and up into the nasopharynx. Even in adults, the amount of continued use of prosthetics is only down to about 20% to 30% at three years. In kids I think it's even less.

For evolving problems, for neurologic problems, the prosthetic devices are certainly worth considering, but they're not a very large part of our practice. So for the other structural problems, then we're looking at surgical intervention. And we can break those off into posterior pharyngeal wall augmentations, we can look at repairing any structural problem with the palate, or we can look at trying to obturate the hole that we see that's back there.

Dr. Alyssa Smith:

Next let's dive a little bit deeper into some of the surgical options that are available.

Dr. J. Paul Willging:

If we look at how to fix problems with the levator musculature of the soft palate, if you have a submucous cleft, the muscles are misoriented, and the goal of the operation is to reestablish the levator sling, to reestablish normal function of the soft palate. You can do that at the time of a cleft repair with an intravelar veloplasty. For most of the submucous clefts that we see, these palates have never been operated before. So you can do a modification of the VY pushback, where you do a von Langenbeck incision, elevate the mucosa off the musculature, separate the levator muscles from the posterior hard palate, swing them posteriorly and realign them in the midline, and then kind of push the soft tissue back to close everything.

More commonly we'll do the Furlow palatoplasty, the double opposing Z palatoplasty. The end result is the exact same. We're taking the levators, which are anteriorly directed, and at the end of the procedure we have reestablished that levator sling, and the functional aspect of the palate is back.

Those two procedures work well in two situations for bpi. One, if you have a relatively small gap, because you're going to gain maybe 20% in length with those procedures. So if you have a huge gap, you're probably not going to have enough tissue to close the velopharyngeal sphincter gap that was

present. If it's a relatively small gap, and the child is young, and I think young is certainly under the age of four, doing the Furlow palatoplasty works very well on those kids.

Once you get to the age of eight to 10, and the child has never had their palate fixed, the patterning of speech is so ingrained that you can change the musculature on the soft palate, but the kids aren't going to use it appropriately to really get maximum benefit. Probably 80% of the kids that we did a soft palate repair on who were over the age of eight ended up getting a pharyngeal flap in addition to their palatal repair. So I tend to kind of limit my submucous collect palate repairs to the younger kids, certainly four and a half and under. The older kids, I talk to the family, we have an option. We can fix the palate and see how it does, or we can just put a pharyngeal flap in, because that's probably what they're going to need.

If we have a gap in the velopharyngeal sphincter area, we kind of look at how big the gap is. The work horse for VPI surgery is the pharyngeal flap. It works very well for almost all cases of VPI where the gap is central. You don't necessarily have to see lateral wall motion at the time of your initial exam to really say that a pharyngeal flap is going to work. The children will not sound any different if the soft palate doesn't hit the back wall, because if the side walls come in they still won't have good closure, so there's no reason to reinforce that lateral wall motion. In the vast majority of cases that we've done pharyngeal flaps who've had no preop lateral wall motion, they've been able to develop lateral wall motion once you put some tissue in there for the side walls to oppose. So they just figure out on their own how to move those side walls.

You could also do a sphincteroplasty. I tend to use those predominantly when I have a lateral gap. If I have a central gap I'm generally going to put a pharyngeal flap in. If the kids are able to close the palate centrally and there's a lateral gap, either one side or both, I'm going to do a sphincter pharyngoplasty. That's taking some longitudinal flaps from the lateral posterior pharyngeal walls, elevate them up, close the donor site, create a recipient bed in the plane of velopharyngeal closure, and then transpose those flaps up into the nasopharynx and pull them to the lateral side. The contralateral side. By doing that you're creating a bump of muscular tissue and mucosa on the back wall of the pharynx, so you're narrowing up from the posterior side anterior, but you're also pulling the lateral walls in so they can close better.

There's a new operation, can't really say new, Dr. Mann up in Grand Rapids, Michigan is a plastic surgeon who's probably published the most on buckle flaps. This is another way of essentially lengthening the palate. These buckle flaps are raised, being posteriorly based, and you're taking probably a flap that's about a centimeter in vertical height, you want to stay away from the parotid duct. But you're going to take some of the oral mucosa and some of the buccinator, and then you elevate them posteriorly on both sides. You then separate the soft palate from the hard palate, and push the soft palate back. Then you take one of your buckle flaps and lay it into the nasopharynx. You take the second flap and put it over the first flap to create the oral mucosal closure. So you're just interposing some tissue to shove the palate back further. This will work fine on the kids who have a normal levator sling, and you just push things back. Has the potential not to cause as much airway obstruction because you're not obturating that nasopharyngeal area.

Kind of the last group that I think about are some of the posterior pharyngeal wall augmentation procedures. In the old days we used to use Teflon. It worked extremely well for small gaps, especially very punctate gaps, where you could see one little spot is causing the leak. We physically can't get medical grade Teflon any longer. When we lost use of Teflon, I've tried a lot of different materials, some of the collagen products, some of the hyaluronic acid products, some of the hydroxyapatite products. We used to use abdominal fat, gluteal fat. All of these materials used to absorb over time. By three

months, certainly by six months, almost all the material was gone. We were ending up doing things again.

Dr. Cofer started using Deflux. It is a hyaluronic acid liquid with some polysaccharide microspheres in it, and this would be injected into the posterior pharyngeal wall, and it seems to be one of these materials where native fibroblasts tend to grow into the matrix, and it seems to be a permanent fix. It was developed more for ureteral reflux, and in some rat studies the material seems to persist for years. I've used it for small gaps, and it seems to work extremely well, and I've not had to go back and revise very many. Sheila Cofer has done her trialogic thesis on the use of this Deflux material for VPI. So I think that's what most people are using at this point.

There is a pharyngeal plexus of veins in the posterior pharyngeal wall. There are no valves in these veins. So just as a matter of precaution, I tend to put these kids on some intravenous antibiotics, generally Rocephin, just to make sure that there's some intracranial coverage, just to make sure that we don't create any intracranial problems by injecting this material. But it seems to work well. It's not FDA approved for use on that site, but again, it seems to work well for that purpose.

Dr. Alyssa Smith:

You mentioned that the pharyngeal flap is kind of the work horse. And I'm going to go ahead and put in a plug for our listeners, that we do have a video up on the Head Mirror website for the pharyngeal flap surgery in case anyone wants to take a look at the steps of that procedure. But Dr. Willging, can you touch on some potential complications of that surgery, and what expected recovery is like?

Dr. J. Paul Willging:

I tell the families that the recovery is very similar to a tonsillectomy, from the pain and discomfort standpoint. It doesn't last quite as long. These kids are pretty much back to normal activities at five days, maybe seven, whereas tonsillectomy, it's seven to 10 days. The most feared of the complications is probably obstructive sleep apnea. We are raising this myomucaneous flap and sewing it into the back edge of the palate. We have to make sure that the lateral ports that we create are of an adequate size. So I put an endotracheal tube through the nostril and use that endotracheal tube as a guide on how far laterally to inset the flap. Generally for children age six and under, I'm going to use a 3.5 endotracheal tube as that sizer. For children that are eight and above I generally use a four endotracheal tube. And depending on the size of the child between six and eight, you'll either go 3.5, or a four.

It's important when you're insetting the flap that you don't wrap the flap around the endotracheal tube. The tube is there so you know where the lateral extent of your flap should be. There should be a little triangle of air anterior to your flap, so you're bringing it out lateral till you touch your endotracheal tube, and that's where you're going to inset it.

If your lateral ports are of appropriate sides, the kids are going to be able to close it with speech and still have an adequate airway for nasal respirations during sleep. I have close to a 5% reoperation rate for obstructive sleep apnea. You can go back in and you can make some little releasing cuts and make those lateral ports bigger. You don't have to revise the entire flap, it's generally a 15-minute procedure. But I tell the families upfront, I've got about a 5% obstructive sleep apnea risk. We can fix it, but that risk is there. We make the ports a certain size, it's up to the child to heal them the size that we made them.

Unfortunately I have another 5% where those velopharyngeal ports are made a certain size, but as the healing process takes place, the ports get bigger. So they have a great airway, but they're still having some velopharyngeal insufficiency through that large port that the child's unable to close. Similar to the back cuts you can make to enlarge the ports, you can make some little mucosal advancement

flaps to narrow the ports as well. And that's again a 15, 20 minute procedure, which is nothing like putting the flap in.

So I have 5%, ports are too small, 5%, ports are two big. But those are probably the biggest complications of the pharyngeal flap: obstructive apnea and persistent leak.

The other thing that we see is where the flap was not raised high enough. Everything has to be in the same plane for a pharyngeal flap to work. If the flap is low at its pedicle, and is coursing up onto the soft palate, it's actually pulling the palate down and inhibiting any natural vertical movement that the flap would've had. So a low flap is actually going to ensure the kid continues to have hypernasality, because the kid's going to leak around the flap. And you can repair that, you can make an incision underneath that low flap, get into that prevertebral fascial plane, and push the flap superiorly again. And then take some mucosal advancement flaps from the ports and advance them down into that donor site you created when you pushed the flap up.

So you can revise a lot of these flap surgeries if things healed improperly for you, but obstructive apnea's the worst, persistent VPI is the second, and that comes in the flavor of, the ports are too big, or the flap was too low.

Dr. Alyssa Smith:

How do you counsel parents on the chance of having a successful surgery?

Dr. J. Paul Willging:

I think you have to be honest with your families, and you have to keep track of your own results. And you also have to define the terms appropriately. With velopharyngeal insufficiency, they have abnormal resonance when you start. And post-operatively I think you have to be looking for normal resonance versus abnormal resonance. And in order for you to get better, you have to know when you do not have normal resonance. Some of those abnormal resonance kids, their results are acceptable. We are listening for hypernasality, we're listening for nasal air emission. We may not have to go back and fix it, but we need to know it wasn't 100% normal. We then have another set of that group who are abnormal residents, who are not acceptable, and those kids we may want to watch and wait, or we may want to reoperate on. If we look at improvement in intelligibility, improvement in their nasal resonance, we're probably in the 99% range.

We can improve everybody, but that should not be our goal. Our goal should be normal resonance when we're done. In some of the syndromal kids, some of the severe apraxia kids, it's unrealistic to expect normal resonance. Improvement is going to make speech therapy easier so they can progress in other areas and become more intelligible as well. So I have about a little over 80% normal resonance at the conclusion of my operations. And again, it's close to 99 with significant improvement. Many families are happy where the kids end up and don't want additional surgery, but I have about a 5% reoperation rate to try to improve the nasalance scores that the kids have.

Dr. Alyssa Smith:

Thinking about followup, what does long-term followup look like for these patients?

Dr. J. Paul Willging:

I get a lot of kids from out of state, so I like to either see the child or have the family call in at about three weeks. Just so I can tell early on how things are going: is there an improvement in speech, are they back to normal diet? I like to get some speech therapy started at that three-week mark. The pain is

gone, the swelling and induration of the tissues is back to normal, and it's a good time to start some therapy to kind of develop the lateral wall motion if they need it, and get the kids back in some articulation therapy, if they need it.

I like to see the kids back in our velopharyngeal insufficiency clinic at three to four months preoperatively. I think that's where the objective assessment of having your speech pathologist listen to your child, and they can tell you it's normal resonance or it's not normal. And if it's not normal, are we okay as far as not having to reoperate, or is this a child that, we need to do something to improve intelligibility?

I don't scope all of the kids. If they have continued hypernasality or a nasal air emission, I tend to scope those kids to identify where the gap is, how big it is, and then we come up with a plan of either watch them for a while, or go back in and reoperate to improve it.

Once the kids have reached normal resonance, we generally don't follow them any more. So we generally tend to see our kids at three to four months. If they are continuing to have some hypernasality and we're following them, we'll generally bring them back a year later. But once we're happy with the result, we really don't bring them back for long-term continued followup.

Dr. Alyssa Smith:

Dr. Willging, thanks so much for joining us today to discuss velopharyngeal insufficiency. Is there anything else you'd like to add?

Dr. J. Paul Willging:

No, I think velopharyngeal insufficiency surgery is something that otolaryngology should have a significant part in. I think when you're going into practice, try to set up a program with a speech pathologist. That is not our specialty area. They will be able to bring a lot more patients into your evaluation because of their contacts out in the community. It also is something that we need to be involved with, because the children who do not have an overt cleft palate are really being mismanaged. They're in the school system, where the problem may not be identified appropriately. The therapist may hear some articulation issues, which may be a compensation strategy the kids have because of their underlying VPI, and it's not going to get better until the VPI is fixed.

So these kids are getting lost if they don't have an overt cleft. If you talk to the dental group that are out there, they're looking in pediatric mouths all day long. Many of them don't realize the significance of a bifid uvula, and they are very interested in looking at the general medical care of their kids. And if you tell them, "If you see a bifid uvula and the child has some abnormal speech, send them to our VPI clinic and we'll see if there's anything substantial going on," the therapists are the same way. If they have a friendly place they can send patients to that they're not sure of what's going on, they can get plugged into the right therapy groups if necessary. And if it's a VPI patient they tend to go off into your surgical treatment pattern.

So I just encourage you, look at the non-cleft VPI. You're not going into competition with plastics, and you're doing a significant service to a lot of these kids. And many of them are going to have an underlying syndrome that's not been identified before, so again, you can have some long-term significant impact on the overall wellbeing of these kids.

Dr. Alyssa Smith:

In summary, velopharyngeal dysfunction is a problem with how the soft palate imposes the posterior pharyngeal wall. This can lead to problems with resonance as well as speech production. Patients will present with hypernasal speech and can have nasal regurgitation of food or liquids as well.

Some causes of VPI include structural problems such as an overt cleft of the secondary palate or submucous cleft palate, as well as neurologic problems such as muscular dystrophy or myasthenia gravis. Patients with velocardiofacial syndrome can have VPI due to abnormal function although there's no structural or neurologic problem. VPI can also be iatrogenic after adenoidectomy.

Flexible speech endoscopy is the gold standard for evaluating VPI, but nasometry and videofluoroscopy can also provide useful information.

Treatment can be with speech therapy prosthetics, as well as speech surgery. Some surgical options available include the pharyngeal flap, which is the work horse of VPI surgery, sphincteroplasty, the buckle flap, and injection pharyngeal augmentation. Additionally, an intravelar veloplasty, or Furlow palatoplasty, can be used for patients with cleft palate.

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 are the three patterns of closure of the velopharyngeal port? The first pattern of closure is coronal closure, and this is the most common. The second most common pattern of closure is circular. And the least common pattern of closure is sagittal.

The second question is, what testing method can provide objective data on the degree of VPI? Nasometry can provide objective measurement of VPI by looking at the ratio of sound energy coming from the nose, compared with sound energy coming from the mouth. This gives a nasalance score. This can be helpful to look at how the child progresses through treatment, whether that be speech therapy or after surgery.

The final question is, what are the two prosthetic options available for patients with VPI? The first option is a palatal lift prosthesis, and this is a good option for patients with a flaccid soft palate. The second option is an obturator with a speech bulb that sits in the velopharyngeal port.

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