

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. Raj Petersson. In this episode, we'll be discussing cleft lip and palate. Thanks for being here Dr. Petersson.

Dr. Raj Petersson:

Thank you so much for having me.

Dr. Alyssa Smith:

So, let's first talk about presentation. How do these patients typically present?

Dr. Raj Petersson:

Most of the time, actually, if it's cleft lip, they will present after prenatal detection. So, ultrasound is pretty good for detecting cleft lips. We'd say about 95% of the time they can be picked up by trans vaginal ultrasound as early as 13 weeks, and probably around 80% through trans abdominal ultrasound, but the resolution isn't quite as good. But most of the time, these are going to get picked up beforehand. Sometimes, in literature, you might see that the rates are 16 to 93% detection with ultrasound, 2D ultrasound. And I think some of that speaks to the experience of the ultrasonographer and knowing what to look for. But most of the cleft lips are going to be diagnosed on ultrasound prenatally.

Cleft palate though is very hard to see. So, it's rare that if it's cleft palate only that you're going to pick it up on ultrasound. A lot of times, when they pick up the cleft lip, they will just call it cleft lip and palate, and just lump it together. And most of the time, they're going to be right because usually we're going to see a palate with a lip just in terms of frequency. So, that's how they're picked up. If it's just cleft palate only, it's usually picked up at birth when they're doing their newborn exam and screening. And then, you'll get consulted at that point.

Dr. Alyssa Smith:

And how about some symptoms that are related to either cleft lip or cleft palate?

Dr. Raj Petersson:

Yeah. So, with cleft lip, most kids are going to feed okay and breathe okay. But those are the things we worry about, is to make sure that they're able to feed whether it's breastfeeding or bottle feeding. And if it's just a cleft lip only, they should be able to breastfeed, but it may be a little bit more difficult depending on the size of the cleft lip. And you may get some nasal airway obstruction. And that comes from the cleft nasal deformity, because you get a septal deviation with that, which we'll talk about a little bit more later too. With a cleft palate, really feeding and airway are the issues.

With a cleft palate, you can't create the suction you need to suck during breastfeeding, or with regular bottles. And so, that will become an issue. And so, they'll need specialized bottles and feeders. And we do worry about airway breathing problems, especially if it's a cleft palate related with other deformities, especially micrognathia.

Dr. Alyssa Smith:

And then, do we know how common this is? And is there any variation in incidents among different ethnicities?

Dr. Raj Petersson:

Yeah, there is. So, in general, a rule of thumb number that people give is cleft lip and palate is one in 1000 births. It's the most common birth defect. So, what I usually go by is that it will be about one 700 births will be cleft lip and palate. Cleft lip only is more rare. And that might be one in 2800 births. Cleft palate only is about one in 700. There is ethnic difference. So, we'd say it's about one in 1000 for Caucasians, two in 1000 in Asian populations, point three in 1000 for African populations. And that's for cleft lip. The cleft palate incidence doesn't seem to differ amongst ethnic groups. In the US, Native American or Alaskan Native populations have a high incidence. So, they might be four point two in 100 for cleft lip, and ten point one in 10,000 for cleft lip and palate.

And then, there are some differences between males and females. So, we'll say, cleft lip and palate, it's about two to one male to female. Cleft palate, however, by itself, is about one to two male to female.

Dr. Alyssa Smith:

So, lets take a second to focus a little bit on embryology, which I know is everyone's favorite. Can you talk a little bit about the normal development and what happens in development that causes a cleft lip or palate to form?

Dr. Raj Petersson:

Yes. So, it's a little bit hard without pictures, but I'll try to describe it as best as I can. But main things to remember are that the lips form between weeks four and seven, and the palate between week six and nine. Around five weeks of gestation, there's ectoderm near the neural plate that folds on itself to form the neural tube. At this point, the embryo is about three milliliters. Then, there are special neural crest cells of ectodermal origin that differentiate to form a special ectomesenchyme. This migrates over around the head area, and participates in the formation of five facial prominences. And this is probably the more important part to remember. These prominences surround the primitive oral cavity. And you have: a frontal nasal prominence, which is midline in the frontal nasal area; two paired maxillary prominences; and two paired mandibular prominences.

So, around the fifth week, the frontal nasal prominence will form these bilateral nasal placodes, which are invaginations of ectodermal thickenings. These will then form nasal pits. And then, this will divide the frontal nasal prominence to medial and lateral nasal processes. At the sixth week, the medial nasal processes will fuse. And these structures give rise to the midline of the nose, the medial upper lip, the philtrum, the incisor teeth, and their primary palate which is the part of the palate anterior to the incisive thrombin. The lateral nasal processes will form the nasal alae and the base. And then, the maxillary processes will form each side of the mouth. They'll grow forward and merge with the medial nasal processes to form the lateral upper lip and the majority of the maxilla and the secondary palate. So, that's the part of the palate behind the incisive thrombin.

And then, the mandibular prominences will fuse in the midline to form the mandible in the lower lip. So, if there's failure to fuse along any one of these aspects, you'll get a cleft.

Dr. Alyssa Smith:

And when we talk about this failure to fuse, is this something that's caused by genetics, or are there environmental factors as well?

Dr. Raj Petersson:

So, we think it's probably a combination of genetics and environmental factors. The etiologies aren't very well known unless we find a syndrome. Most of the time, we won't find the cause. So, we say 50 to 60% of the time it might be sporadic. And that might just mean that we haven't identified the gene yet. But we usually think of it as an interplay of some genetics and environmental factors. So, certain environmental factors that we know of that may contribute to clefting are: smoking during pregnancy, diabetes, certain medications, especially anti seizure medications like topiramate and valproic acid, steroids, retinoids, Vitamin A, which is why we worry about things like Accutane and Lithium. Fetal alcohol exposure may lead to it, or maybe certain infectious processes.

Dr. Alyssa Smith:

And so, you've already mentioned kind of one term, a secondary cleft palate. But I think it might be helpful to define a lot of the terms that we use when describing these defects. So, can you take a second to talk about incomplete and complete defects, and then these primary and secondary defects as well?

Dr. Raj Petersson:

So, if we look at lips, we kind of break it down into some broad categories. So, we decide if it's unilateral versus bilateral, right versus left. Is it incomplete, which means that it's only involving a portion of the vertical height of the lip and doesn't extend all the way through? And this could be as small as a little defect in the vermilion border, or all under the skin such as in the microform cleft lip. A complete cleft lip will go through the entire vertical height of the upper lip and maybe into the nose, into the nasal seal. And that will often be associated with the cleft and the alveolus. After we look at the lip, we look at the alveolus to see if there's a cleft there. And that can be anything from a notch to going all the way through the alveolar ridge.

In terms of the palate, it can involve the primary or secondary palate, which means it can be complete or incomplete. And there's all different ways to look at this. And so, in a lot of my training, we just kind of described it as: Is it soft palate? Is it narrow? Is it wide? Does it come all the way to the alveolus?

But one helpful classification could be the Veau Classification system, which divides them into four groups. And so, group one would be soft palate only. Group two is hard and soft palate to the incisive thrombin. Group three is a complete unilateral cleft palate that involves the soft hard palate alveolus, and usually with the lip. And then, Group four is a complete bilateral cleft of the soft and hard palate, which may or may not include the lip, but often does, as well as the alveolus. So, a bifid uvula would be like the least severe form of a cleft palate. And a submucous cleft palate is the mucosa is intact, but the muscle doesn't come all the way across. And the muscle we're talking about is the veli palatini.

Dr. Alyssa Smith:

So now that we have that in mind, let's talk a little bit about work up. And when you're evaluating a patient with a cleft lip and/or palate, what are some important questions that you ask about when gathering a history?

Dr. Raj Petersson:

In terms of the work up... Whenever I think about work up and history and physical, you always think about what you're going to ask for any patient that you see in a pediatric ENT clinic. But we kind of just start from... In terms of the cleft lip or palate history in particular, we really want to find out: Are they having problems feeding? Are they having problems breathing?

Especially with a cleft palate, we know they're going to have more problems feeding. So, we ask questions about weight gain. What are they doing for feed? How are they feeding the baby? Are they trying to breastfeed or are they bottle feeding? Breast milk or formula? And we try to assess their overall health and make sure they're gaining weight. We always ask about breathing. Again, with severe cleft nasal deformity, they may have some trouble breathing, as babies are obligate nasal breathers up until three or four months of life, if they have a small GI or a micrognathia. They may have trouble breathing from tongue based obstruction. We always ask about the ears. And so I ask if they passed the newborn hearing screen or not. Then, we always ask about family history, so if there's any family history of clefting, if there's family history of hearing loss or syndromes that could be associated.

We look for health of other organ systems, knowing that cleft lips and palates may be associated with other things that may lead to a diagnosis of a genetic syndrome. So, I ask if there's been any heart issues, ear issues, any other skeletal issues, or genital urinary issues. And then, we always give the parents opportunities to ask ample questions, because they're going to come in with a lot of questions. Now, in terms of the exam... And we do a complete head and neck exam. So, at first, you kind of look at general. How are they looking? Do they look like they are gaining weight? Do they look very thin, failure to thrive-ish. I look at their breathing. Do they have any stridor or stertor? Do they have any retractions. Then, we look at the ears, see if there's affusions. Because if there's cleft palate, they very well likely may have affusions. Look at their overall facial features to see if you see any signs of dysmorphism, which can be really hard to tell in a newborn. But things may pop up as they grow and get older.

Then, we look at the lip and palate, and try to put it into one of these categories that we've already talked about, to understand and get an idea of how severe the problems are.

Dr. Alyssa Smith:

So, when these patients come in for evaluation, is there any other imaging that you obtain? Or do you get photographs of them? What else is involved in the work up?

Dr. Raj Petersson:

Yeah. So, we get photographs. We do refer all of our patients to genetics for evaluation. Part of this is to help get a proper diagnosis if it is going to be syndromic, also for counseling. They can talk about dental recurrence risks. They can have more in-depth conversations about what may or may not have caused this. Some common syndromes that are associated with clefting are Van der Woude... If you see lower lip pits as well, it's very likely to be Van der Woude Syndrome. I've talked about micrognathia, and so we've talked about Pierre Robin sequence, which is micrognathia, or a small jaw, which causes glossoptosis, or the tongue to fall back, which then causes failure or effusion of the palatal shelves. And you can usually get a wide U-shaped cleft palate with that.

22q11 syndromes, or deletions, are important in clefting as well, especially if you find just an isolates submucous cleft palate. We will send all of those kids for genetics evaluation to rule out any 22q11 deletions. But other syndromes... There's more than 300 syndromes that have been described. But ones that you may more commonly see are Treacher Collins, or Stickler, or Waardenburg, Klippel-Feil, trisomies 13 and 18. So those are all things to look for, and why all of these other questions when you take their history are important about other organ systems.

Dr. Alyssa Smith:

So moving on to treatment of these patients, let's first touch on the timeline for surgical intervention. When are these children candidates for surgery?

Dr. Raj Petersson:

So, in general, we repair the cleft lips between three and six months. Sometimes, it may be as early as two months. If they're a bilateral lip, we may wait until six months, because we're going to cause more airway obstruction when we close their lips. So, that's about the timeline of the lips. And in general, for a palate repair, it's going to be between 10 and 12 months. If it's a soft palate only, we might creep down to eight months. If it's [inaudible 00:14:24] with Pierre Robin sequence, and we're more worried about the airway, we may wait until they're over a year to repair it.

Dr. Alyssa Smith:

And so, with that in mind, thinking about the time that it takes to actually get to the point where a patient would be a candidate for surgery, I think a lot of us have heard about Nasoalveolar Molding. Can you touch a little bit on the roll of that?

Dr. Raj Petersson:

Yeah. So, that can be helpful, especially if you have a wide defect. But Nasal Alveolar Molding is an appliance that's made by dental specialists that mold things into the place and shape that you want. So, it will help mold the nose with a wire and acrylic stint. And it's attached to an intra oral denture. So, it will mold the nasal cartilages into a more desired shape. It can help with the premaxilla in bringing the alveolar ridges together. It can reduce the severity of the defect before surgery to hopefully optimize your outcome. And so, a good role for these may be wider clefts, or bilateral cleft lips and palates, if you have a prominent premaxilla.

The issue with Nasoalveolar Molding is that requires weekly visits to the dental team to adjust it. Because the baby is growing, but also we're changing the anatomy. So, it needs weekly adjustments. And so, this can be difficult for most families. And it's something that we don't do routinely at my institution, nor did we do it routinely where I did fellowship, because of difficulty to get patients to be able to come in to do this. A lot of our patients are coming from far away. And they've got a newborn. So, it's a lot of appointments, and they have a lot of other appointments anyways. Other downsides are potentially that the nasal stint may cause nasal obstruction, which can make it hard to breathe, or these can dislodge and potentially cause an airway obstruction. So, there are centers who use these a lot, but they may have easier access to their dental teams, patients who are able to get there. But there's a lot of cost and time involved, so it's not something that we use commonly in our practice.

Dr. Alyssa Smith:

And then, I know I've heard about lip taping. Is that something that you commonly use? And what are the benefits and drawbacks of that?

Dr. Raj Petersson:

Yeah. So, that's something we can use, and that's something that is much easier to do and costs much less. And so, the time you would use that potentially is if you've got a really wide lip. And it's more rudimentary than the Nasoalveolar Molding, but you basically are taping from the cheeks, across the lip, to bring the segments together. And the parents can be taught how to do this. There's not a whole lot of downside other than potential skin irritation from the taping. And there's potential upside that you may be able to narrow your segments to make the surgery easier. But even with that, we find that it's not something that families can comply with very well and keep up with, for the few months that they have to do it to get to surgery.

Dr. Alyssa Smith:

So thinking about the surgery and the goals of surgery, I know that these surgeries would be a lot easier if we just thought about them as closing the gap, or closing the hole. But obviously, it's a lot more complex than that. Can you talk about... to the goals of surgery?

Dr. Raj Petersson:

Yeah. So, the goals of surgery, in the most simple terms, are to fix the hole and restore the function. But what that means is fixing the whole in an aesthetically acceptable manner, worrying about the nasal airway, restoring function, which means muscular function. So, for lips, it's going to be making sure you bring the orbicularis oris muscle together. For palates, it's going to be bringing the levator veli palatini muscle together in the soft palate. And so, those are things that we try to do. In doing that, we also want to have things heal in the best way possible, with unobtrusive scars, good symmetry between the normal side and the cleft side.

Dr. Alyssa Smith:

And so, you mentioned bringing some of those muscles together. Can you talk a little bit about the different patterns of the way the muscles lay down for a patient with a cleft lip or palate?

Dr. Raj Petersson:

Yeah. So, with the cleft lip, the orbicularis oris muscle, instead of forming a sphincter and going across the lip, will have abnormal attachments to the nasal base. So, instead of coming across where the cleft is, the fibers are going to swing superiorly and attach to the nasal base. So, in repairing them, we need to release all those abnormal attachments, and bring them across the midline. In a cleft palate, the levator veli palatini, instead of going across the palate and forming a sling, are going to attach anteriorly to the hard palate. And so, when we're repairing the palate, we need to make sure we release all those muscle fibers and bring them across midline to try to re-create that sling.

Dr. Alyssa Smith:

And so, let's first focus on repairing a cleft lip. What are some surgical approaches that we can use?

Dr. Raj Petersson:

So, there's lots of different ways. But they fall into two broad groups that are typically used. And that's going to be a group of techniques called rotation advancement type techniques and a group of techniques called triangle techniques. With the rotation advancement type technique... And the most common one here that most people will have heard of is called the Malar Technique. There's also Molar Technique, among others. With that rotation advancement, we're trying to take the medial segment, which is the one coming from the normal side... So, take that medial segment and rotate it inferiorly. And then, the lateral segment will come in, advance medially, and be placed in a way to kind of hold that medial segment that we rotated inferiorly down, so that we achieve lengthening of the lip.

The major problem, other than the hole itself, is that the cleft side lip is going to be shorter than the non cleft side. And so, the object here is to even out the length. So, if we just closed it, it would stay short and would still look like there's a defect. So, that's the idea there with the rotation advancement. With the triangle techniques, we're also trying to achieve that length. But here, we use a triangle on the lateral side to insert onto the medial side to achieve that length. And there's various placements of the triangles, but usually it's going to be just above the cutaneous roll of the lip. The Malar technique is a

very common technique that a lot of surgeons use. And this involves mostly lining up incisions along the philtrum column. But, on the plus side, to achieve that length, it's going to curve towards the non cleft side at the upper aspect of the philtrum, below the base of the nose, which is where you're going to get your rotation from. And then, you're going to be designing a flap on the lateral side to kind of fill into that defect.

The Molar Technique is a modification of this, where the back cut comes up into the columella of the nose. You don't have any scars that go across the philtrum, but up into the columella. So, it may hide better up there. And then, with the triangle techniques, the only incision that you may have going across the philtrum is a small back cut for where you're going to insert that triangle. The techniques that fall under this category are, for example, Tennison-Randall, or the Fisher technique, which was ascribed in 2005, which is the one that I've started using now.

Dr. Alyssa Smith:

And so, do you kind of pick your approach for the different lip and the deformities that that lip has, or does every surgeon kind of have their favorite technique that they use?

Dr. Raj Petersson:

Yeah. So, everybody does this a little bit different. So, some surgeons will just have one technique and use that for everything. In my training in fellowship, we were taught various techniques for various types of lips. So, for me, in general, for a microform lip, I might use the Malar technique because I don't have to make that big of a back cut into the philtrum, so it's not that big of a scar there. For a narrow unilateral lip repair, I might use the Millard technique or the Fischer technique. And then, for a wider cleft lip, I might use a Fisher technique, or the Tennison-Randall triangle technique. In my hands, I just find it easier to get all the length I need in a wide short lip with a triangle technique over the Millard technique.

A lot of times, with the Millard technique, I find if you have a big height discrepancy that, as it scars, it'll pull that lip up. Whereas, using concepts of triangles kind of buffer against that shortening better.

Dr. Alyssa Smith:

And are these the same techniques that you would use for a bilateral lip?

Dr. Raj Petersson:

No. So, for a bilateral cleft lip, the technique is a little bit different, because you don't have anywhere on the medial segment to get a lot of length from. So, in that medial skin lip segment in front of the premaxilla, the tissue that you have is all the tissue that you're going to get. So, with a bilateral cleft lip, we use a method called the straight line closure of Veau. Mulliken out of Boston has made modification to this, which is what I use. But here, the idea is to bring those lateral segments together. With the medial segment, you're going to create a new philtrum there. There's no muscle across that middle segment in front of the premaxilla. And so, ideally, you need to bring the muscle from the side all the way across the middle.

Some concepts here are that when you bring those lateral segments across, as the child grows, you're going to get stretching of what's left in that philtrum. So, we actually make the philtrum segments, or flaps, that we want to use much narrower than what they're going to be because we know they're going to be wide and out. So, we know the normal philtrum becomes about a centimeter. But we might make that medial bit, and whittle that down to five milliliters or so, so that it's pretty narrow

knowing that it's going to stretch. There are different challenges here. And I would recommend, with all of the techniques I mentioned, just going online and finding the pictures of them to really be able to visualize it in your head what they are.

Dr. Alyssa Smith:

And so, I know when I've kind of thought about this and tried to learn it, I have seen a lot of pictures that have these skin markings on them for the different points and the anatomy that we need to have in mind when performing the procedures. Are these skin markings ones that are also used in surgery as well?

Dr. Raj Petersson:

Yeah. So, the skin markings are really important. So... Like right now, I'm looking at... myself at the markings for the Fisher technique. And there's 25 markings. So, sometimes I'll even bring the pictures into surgery with me, or take pictures of the kids and mark it out on the picture and bring it with me for reference, just so... These aren't surgeries that we're doing 10 of every day. So, it's just a good reminder to make sure that you're getting them, because the markings are the most important part in surgery. The rest is cutting and sewing which, as surgeons, we all can do. So, if we don't get the markings correct to begin with, we're not going to get the result we want at the end.

But some key points to mark are the border of the red lip, the vermilion, and the cutaneous junction. So, the cutaneous roll is a really important marking point. So, we do mark those in fine markers. And then, we'll methylene blue any points that we want to make sure don't get erased during the surgery. So, the points that we mark are... so, the trough of Cupid's bow on the lip. On the normal side, we mark the peak of the bow. And then, we measure that, and then create a corresponding mark on the cleft side, because you have to figure out where that point's going to be and you want it to be symmetric. And that point, I will mark with methylene blue so it doesn't get erased, so that I know where to mark it compared to the other side.

Then, on the lateral side... So, on the cleft segment, we'll also mark the border of the cutaneous Vermilion junction so that you'll know that those two points have to come together. So those may be the only two points that I mark in methylene blue. Everything else I usually mark in marker. And once those incisions are made, you've kind of made all of your cuts. But those are two important points to remember. Then there's nuances that come with depending on whatever repair you use, where you need to bring the nasal seal together, depending on what [inaudible 00:27:40] looks like. So, you may make some [inaudible 00:27:42] and measure those areas.

I often put a triangle in the Vermilion part of the lip, because you're going to have some deficiency in bulk on the medial aspect. And so, I'll usually make a back cut on the medial aspect, and mark out a triangle on the lateral aspect. We always want to make sure that we line up the dry and wet junction of the lip together. So, everything you're doing is to try to not make any mismatches that are going to be easily picked up by somebody looking at it.

Dr. Alyssa Smith:

And then, before we move on to the cleft palate repair, are there any technical pearls or common pitfalls that you see with cleft lip repair?

Dr. Raj Petersson:

So, I think the big things to remember are achieving length on the short cleft side to match the normal non cleft side. You want to hide scars in normal anatomic sub units as much as possible. So, the philtral



ridge is an area. The nasal seal is an area. So, you don't want to be making any scars that go across sub units. That's going to be easy to see. Careful tissue handling is really important, especially in baby skin, so it easily bruises. And that will help in healing as well. Really bringing the orbicularis oris muscle together in the midline, so releasing all the attachments from the nasal base and making sure that those are closed separately in the midline, lining up their dry and wet vermilion, and then lining up that cutaneous roll. So, those are going to be sort of the over riding principles no matter what technique you use.

Dr. Alyssa Smith:

So, let's move on to cleft palate. What are the approaches for cleft palate repair?

Dr. Raj Petersson:

So, the cleft palate repair approach is going to depend on what you have for your cleft palate. So, you're going to use potentially different techniques for completes versus incompletes. General principles, again, are going to be fixing the hole and restoring function. We always close it in two layers. So, there's a nasal layer and oral layer, really is three layers because there's a muscle layer too that we worry about. We want tension free closure, just like we do with any surgery that we do. And then, you want to know is there a way to also lengthen that palate to help with speech.

I'm going to just go through some broad categories of repairs. And again, I definitely encourage looking these up for pictures to help. But one repair is called a two flat repair, which is pretty common. And this a good one for complete cleft palates that come all the way up through the primary palate. What it involves is having oral mucosal layers that are based off the greater palatine artery. And very important when we're doing dissection to be careful around the expected area of the artery, which is going to be just medial to the second molar, or where the second molar should be. That is going to be the blood supply for these flaps. So we're very careful around those. The oral layers are dissected off the hard palate. And you dissect them with the muscle layer off the soft palate. And as we're doing that, we also release the abnormal attachments of the muscle from the hard palate edges, so that that can all come across on the midline.

And then, the nasal layers are dissected separately. And so, you close it with the nasal layer as a separate layer. The muscle, there's different things that people can do with it. You can dissect all the muscle out and close the muscle separately. Sometimes, people will just close it all together with the oral layer. The key is to get that muscle across midline. The big thing with this procedure by itself is it does not lengthen your palate at all. And when you have a cleft palate and you close it, it's generally going to be short.

So, we do start thinking about are there ways to lengthen that palate. So then, you have another technique called a V-to-Y pushback, which is better for an incomplete cleft palate, one that doesn't come into the primary palate, because your incisions won't come all the way up to there. But same general ideas where you dissect all the mucosa off the hard palate, separate the muscle. But what you do here is, in the process of bring the flaps to the midline, you're actually pushing it back. And because you have intact mucosa anteriorly, you are pushing it aback and attaching to that mucosa.

There's another technique called a von Langenbeck technique. This is for incomplete cleft palates. And if I were to use this technique, it would be for ones that don't come up too far into the hard palate. But here, you're not making any incisions in the midline anteriorly, so you have lateral incisions. And you're doing a lot of blind dissection, so you have to be really careful around the greater palatine artery. But it makes the healing part easier, because you have less raw surfaces. This also does not lead to any lengthening. And then, there's a technique called the Furlow Palatoplasty, or double opposing z-

plasty. And this is a technique utilized on the soft palate which will actually lengthen the palate. So, you design z-plasty flaps with the oral layer and the nasal layer. And again, it's really hard to describe without looking at pictures, or even for me just seeing the surgery for the first time is when things became very clear. But the general concepts are z-plasties lengthen incisions. And we re-orient the levator veli palatini muscle with this technique to be in the more correct anatomic position.

So, the flaps are designed to achieve those two goals. The left oral flap is a mucosa and muscle flap, and designed in a way to [inaudible 00:33:28] the muscle posteriorly off the abnormal attachments. And the right oral flap is a mucosa and sub mucosa flap. And the nasal flaps are oriented in the opposite direction. And the left one is mucosa and sub mucosa. And the right one is mucosa and muscle. And so, when you bring those two z-plasty flaps, the muscle comes across midline and the two z's make the palate longer. And this, you can combine with any of the other techniques I just talked about, if you want to add it to your complete cleft palate repair.

Dr. Alyssa Smith:

So basically you could use the other techniques to get a good closure, and then get the length with the Furlow technique.

Dr. Raj Petersson:

And the Furlow technique is a really good one for soft palate cleft only, or a submucous cleft palate.

Dr. Alyssa Smith:

And so, thinking about one of the feared complications of cleft palate repair, a fistula, what are some things that we should have in our head to reduce that risk as much as possible?

Dr. Raj Petersson:

Right. So, the biggest thing is tension free closure. And the most common place that you're going to get a fistula is at the junction of the hard and soft palate, because that's where the most tension is. And the reason the most tension is there is because that's about where the pedicle is. This isn't so much of a problem in a narrow palate, but becomes more of a problem the wider it gets. And so, there are certain things that we do to help with that tension free closure. So, some surgeons may strip the muscle off the hamulus, so that we're not attached there. To be able to bring those flaps across the midline, you really have to do a lot of dissection around the greater palatine artery pedicle. And this can get scary because you're always worried about harming or injuring the vessel. But it can be done. And there is actually pretty good amount of periosteum around it.

So, a lot of times, you really just have to keep dissecting that vessel out so that you have a lot more mobility. The nasal flaps are generally closed without any tension. So, there's a lot of laxity in the nasal mucosa, and the tension isn't much of a problem there. But it can be in those oral flaps. And so, things that I do are either strip the muscle off the hamulus... Some people may intra fracture the hamulus on both sides to be able to get those flaps closer. And then, I think you really do need to do some careful but good dissection of the greater palatine arteries to be able to get that mobility. And you don't want to throw any stitches until you've played with those flaps and brought them across the midline, and you can see that they're tension free. Some people will place a layer AlloDerm. I'm not sure if this helps or not. But potentially, when it's really wide, if you need an extra layer, that may be helpful. But it's not always required. It's not something that I use very often.

Dr. Alyssa Smith:

So, let's talk a little bit about the typical post operative regimen. And if you can touch on whether or not these children need antibiotics or steroids, and even what the feeding schedule looks like post-op.

Dr. Raj Petersson:

Most of our kids are going to get a dose of antibiotics in the operating room. And I usually give them a dose of steroids. We use Decadron, 0.5 mgs per kg, especially for the palat repairs because they're going to have a lot of swelling and potential airway obstruction. With the palate repair also just important to remember in terms of airway is that mouth gag that we use, which is called a Dingman Mouth Gag, just like when we do tonsils and adenoids, puts a lot of pressure onto the tongue. So, I do release that gag every 20 minutes just to let the tongue re-profuse. And doing this can help you avoid having to use a tongue stitch or other things that you may hear about. So, I think that definitely helps.

So, we do a dose of steroids in the OR. Antibiotics post-op I typically don't do. Maybe we might do 24 hours if we're worried about infection. In terms of feeding, if it's cleft lip only, they can typically just go back to their normal feeding. Of course, they're going to be sore. And so, you treat their pain with Tylenol. But they can typically go back to normal feeding. They may have some airway obstruction. We didn't mention it before, but a lot of times we do do a primary tip rhinoplasty during the lip repair. So, they may have some swelling inside their nose. And so, you just need to watch for that and see if it's affecting their feeding.

With a cleft palate, they can typically go back to the way they were feeding before. The idea situation is that they're off any of their bottles and on to a free flowing sippy cup. But, at this point, a lot of surgeons will allow them to just go back to their specialized feeders as well, but they're going to hurt. So, good pain management and I just alternate Tylenol and ibuprofen at their maximum per weight doses every three hours. We really try to avoid narcotics, can usually do so. But you have to really watch their feeding. And so, it may be even a temporary period of time where you're feeding them through a syringe, or a spoon, or just trying to get it into them. And it can be a lot of work for the family, especially in those first few days.

And then, in terms of wound care, there's not a whole lot of wound care to do. For our cleft lip patients, I typically put Dermabond on them to kind of keep them water proof, because they're still going to have snot and mucous and saliva. And so, that is definitely helpful with our cleft palate kids. We also tell them nothing crunchy, no puffs that they might be eating, so just soft mushy things, nothing grainy, so not rice cereal. We don't want anything that can get stuck in the suture lines. I usually have them rinse with water after every feeding.

Dr. Alyssa Smith:

And so, I'm thinking about the follow-up schedule. What does that look like for these patients?

Dr. Raj Petersson:

Usually, I'm going to see kids back about a month after surgery just to make sure everything is healed well. After that, a follow up is going to depend on what their needs are. And this is where our multi disciplinary cleft teams come in. So, we will follow a lot of these kids through the teams. So, one thing I didn't mention before is that, in general, at the time of the cleft palate repair, we're also placing ear tubes because we know all these kids have eustachian tube dysfunction from their eustachian tubes not working properly because of the cleft in their palate and the gap in the levator veli palatini. So, they all get ear tubes.

And so, we check those in the post-op check. And then, we check the ear tubes every six months like we would for most kids with ear tubes. Sometimes that may coincide with team visits that they're

getting anyways as well. Sort of moving forward from them, things we're monitoring for during their team visits are feeding and speech when they're old enough to produce speech. We have a dental team. As part of the teams, they're looking at dental occlusion when teeth start coming in, health of their teeth, any supernumerary teeth, or carries or anything like that. We have a team psychologist who's there to support families, but then also, as these kids get older, if they have any needs. We have genetics on the team so they can give families updates if anything is coming through in regards to their testing or their certain diagnosis. We have... On our team, the clefts are done by either myself, so ENT, or plastic surgery. And so, we're both on the team looking at our respective things.

We have oral surgeons on the team when the kids are older and they may need some oral surgery care. We have a team coordinator who helps kind of manage and coordinate all of these things. These are all important, I think, to the success of what we're doing in coordinating procedures and surgeries, making sure people are on the same page, and we're all kind of working together to not interfere with anybody else's treatment plans, or contribute to their treatment plans.

Dr. Alyssa Smith:

Well, it sounds like multi disciplinary care provides a lot of benefits for these patients. Dr. Petersson, thank you again for joining us. Is there anything else you'd like to add?

Dr. Raj Petersson:

I would definitely encourage kind of going and looking at pictures of the techniques I talked about. They're going to be the most common ones out there. And I think that is what helps, kind of looking at the picture while you're hearing about it. So, I would recommend doing that. There's a lot involved in the care of these children. And so, the team care is very important so that everybody can bring their own expertise to the table. We have a timeline for these kids all the way to adulthood, and it's helpful to show them a chart of sort of things to expect as they go along. So, repairing the cleft lip and the palate is the first part of it. Twenty five percent of the time, after we repair the palate, they may have velopharyngeal insufficiency which might require a second surgery on the palate. And so, those speech evaluations are really important.

And then, they may need revisions along the way. At some point, they're going to need a bone graft. If they have an [inaudible 00:42:19] at the oral surgeons due, they may need revisions of their lip or scar revisions. And then, they may have other maxillofacial issues that need addressing. So, they may have maxillary hypoplasia. And so that's where all of these people come in. And then, sort of towards the end, as they're getting fully grown, one of their last steps may be their final cleft rhinoplasty where we take care of their septal deviations, and their nasal deformities. That's one thing that we didn't talk about. And things to remember are that the cleft ala is inferior and lateral to the contra lateral side. The ala rests on an underdeveloped premaxilla. The lower lateral cartilage on the cleft side is underdeveloped and weakened, and the dome is lowered. The columella is fore shortened. And the septum and columella deviate towards the cleft side. But the columella base is going to deviate towards the non cleft side. So, I just wanted to get that in too.

Dr. Alyssa Smith:

Awesome. Again, thanks so much for joining.

Dr. Raj Petersson:

You're welcome.

Dr. Alyssa Smith:

In summary, cleft lip is the most common birth defect and in general occurs in one in 1000 births. Clefts can be unilateral or bilateral, complete or incomplete, and can involve the lip, palate, or both. The Veau system can be used to classify cleft palates. During embryo genesis, there are five important facial prominences: the frontal nasal prominence, two paired maxillary prominences, and two paired mandibular prominences. The front or nasal prominence will develop nasal placodes, which will divide the lateral and medial nasal processes. Failure of fusion of these processes and prominences will lead to the development of a cleft. Many children with cleft lip or palate are diagnosed on prenatal ultrasound, although this is less sensitive for isolated cleft palate. In general, cleft lips are repaired between ages three to six months, while cleft palates are repaired around 10 to 12 months.

The two groups of surgical approaches for a unilateral cleft lip include a rotation advancement technique, which includes the Mallard technique, and then the triangle technique. For a bilateral lip, a straight line closure is used. For cleft palate, a tension free three layer closure is important, where there is closure of the nasal, muscle, and oral layers. This helps prevent development of a fistula. Techniques used in cleft palate repair include the two flap repair for complete cleft palate, the V-to-y pushback and von Langenbeck techniques, which are good for incomplete cleft palates, and the Furlow technique, which is designed to lengthen the soft palate and can be combined with any of the techniques used for hard palate repair.

Finally, multi disciplinary teams are helpful in providing the complex comprehensive care that these patients require. 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 differentiates the primary and secondary palates?

The incisive foramen separates the primary and secondary palates, with the primary palate being anterior to the foramen and the secondary being posterior.

The second question is: What are the four classifications of cleft palate in the Veau classification system?

So, group one is a midline cleft of the soft palate with an intact hard palate. Veau group two is a midline cleft of the soft palate and secondary hard palate with an intact primary palate. Veau group three is a cleft of the soft palate extending unilaterally through the secondary hard palate and through the primary hard palate and alveolus. And then, Veau group four is a cleft of the soft palate, hard palate, up the incisive foramen, and then bilaterally through the primary hard palate and alveolus on each side.

The third and final question is: What is the general timing for surgical repair for a cleft lip and a cleft palate?

So, in general, cleft lips are repaired between ages three to six months. And then cleft palates are repaired around 10 to 12 months. Thanks for listening and we'll see you next time.