JEFREY SIMONS: Well I'm delighted to be here, and we spend the next hour talking about the evaluation of noisy breathing in infants and children. And as David mentioned, there's a lot to cover, so I want everybody to leave with a general approach to these issues, and to take some key points home with you. There's a lot more information on the slides that you can refer back to later.

I also want to encourage all of you to send email questions in, and we'll try to get as many of them as we can as we go along this evening. I have no conflicts of interest to disclose. The main objectives of this discussion are to understand a rational approach to the evaluation and management of infants and children with noisy breathing, and to strictly describe common causes of stridor and airway abnormalities in the pediatric population.

When we talk about noisy breathing in children, it's a common complaint. Lots of kids present to the doctor or to the emergency room with noisy breathing. It implies that there's a perturbation of airflow at some level, at some site. There are many different potential sites and a variety of potential noises. The goal is to determine the exact site of obstruction, the cause, and then to treat it appropriately. That's the overall goal.

The pediatric airway differs greatly from that of the adults, and some of these things are key issues for people taking care of children, particularly in otolaryngology who are taking care of children to know about. The airway has a smaller diameter in children, in infants, particularly. Therefore, minimal edema can lead to critical airway distress. So the normal neonatal subglottis is only four to five milimeters in diameter, compared to 12 millimeters in an adult. The cartilage is more pliable, so there can be collapse of the cartilage, which is malacia. There could be developmental issues related to feeding and swallowing which can contribute to airway symptoms, and conversely, airway symptoms can also lead to more problems with feedings and swallowing, so that can go both ways.

Also, congenital anomalies and hemangiomas tend to be more common, unlike adults, where you think about malignancy and autoimmune diseases as a cause of swelling in the airway. The other issue that we have, which is challenging in taking care of children and infants, is that they often can't describe their symptoms. So we have to be a detective to figure out what's going on.

We're going to talk about some different types of noisy breathing, and these are important

terms that are used commonly by pediatric otolaryngologists and general otolaryngologists to talk about different types of noisy breathing. Snoring tends to be produced by the soft palette and uvula vibrating against either your nasal or laryngeal wall. And that is similar to stridor but a little bit different. We usually think about stridor as the sound produced by the base of tongue in the hypopharynx. It's a harsh, rattling sound. Usually it's heard, but not always, during sleep. And that's because there's decreased air, muscle tone, and gravity. Stertor tends to be inhibitory. So we sometimes see patients in the office that have stertor during the daytime also, not just at night. And we have to think about what that cause is.

And that can be contrasted with stridor, which is a harsh musical sound produced by the turbulent airflow through the upper airway, the larynx and the trachea. Stridor can be inspiratory, expiratory, or both. And the type of stridor that you have can help direct the differential diagnosis for the cause.

And there's also wheezing. Wheezing is a lower airway sound. Tends to be made by the walls of a narrow airway vibrating against each other. It's usually expiratory. Now patients will sometimes come into the office and the parents will describe, they'll say, my child's wheezing. And you have to really take a careful history and talk to them because sometimes they're really talking about stridor and not wheezing.

DAVID: I think Chavalier Jackson said that, "All that wheezes is not asthma" or something to that effect.

JEFF SIMONS: I think that's true. "All that wheezes is not asthma". But there's actually a lot of other causes of wheezing, but you also have to make sure whether it is wheezing or a different type of sound-- stridor or stertor.

So we'll talk a little bit about the pathophysiology. Some of these ideas are important to help us understand the proper work up of children with noisy breathing. The neonatal subglottis as I mentioned, is small. It's normally only four to five millimeters in diameter. A preemie's subglottis is even smaller-- three and a half millimeters in diameter, compared to an adult with a 12 millimeter normal subglottic airway.

So airflow through a tube, through the subglottis, is governed by Purcell's law, which says that resistance is related to the inverse radius of the fourth power--

DAVID: fourth power--

JEFF SIMONS: --so the fourth power. So small changes in diameter, or in radius, can lead to significant changes in airway resistance. So the airway resistance increases exponentially as the subglottic diameter narrows. If you take an infant's subglottic diameter, which the normal subglottis is four to five millimeters-- and you narrowed that diameter by one millimeter, it decreases the cross-sectional area 75%. Pretty significant.

DAVID: 25% though, still left.

JEFF SIMONS: 25% is still left. That's definitely true. But by doing that, the resistance increases 16-fold because of Purcell's law.

DAVID: That would go from 100% to 25%. Resistance goes up by 16 times.

JEFF SIMONS: 16 times, that's right. And if you compare that with an adult, where if you narrow the diameter of an adult airway by one millimeter, the cross-sectional area only decreases 30%. And resistance changes only two-fold. Really not such a difference.

So for example, if an infant gets a viral infection of the upper airway, just an upper respiratory infection, it can cause some edema in the airway, in the subglottis. And it can cause croup because such an increase in resistance causes turbulent airflow. An adult that gets the same, exact viral infection may get the same amount of edema, but comparatively, there's much less change in resistance and may have no airway symptoms at all.

DAVID: So that's why pertussis is so much more of a deadly disease in infants than an older individuals.

JEFF SIMONS: That's true and in the past there was a lot of other infectious diseases that were much more dangerous in children than in adults because of the size of the airway. Even diphtheria is another example. So we have to ask ourselves, can the site of airway obstruction be localized by the sound quality? Well, somewhat yes.

You have to be careful about snoring versus stertor because although they're different, there's somewhat overlapping terms. Snoring tends to come more from the nasal pharynx and the palate, but it certainly can come from the tongue base, tonsils and uvula as well. Stertor tends to more come from the base of tongue, tonsils, and hypopharynx. Again those two are sort of overlapping. But they can be differentiated from stridor, which tends to be a noise coming from the larynx or trachea. Or wheezing, which tends to come from the peripheral lower airway.

Another question we can ask ourselves is, can the site of airway obstruction be localized by the phase of respiration? Again, sometimes we can get some good clues. Inspiratory noise tends to come from the glottis and above. It tends to be extrathoracic.

So snoring or stertor, you often think about pharyngeal soft tissue collapse. Stridor, you think about a collapse or some kind of obstruction involving the supraglottic larynx or the vocal folds. Expiratory noise, which can also be grunting in some children, you think about interthoracic trachea-bronchial disorders. And if there is biphasic noise, it's also often from a fixed obstruction, usually involving the subglottis or trachea. Does that all make sense?

DAVID: Absolutely.

JEFF SIMONS: It's important to think about the differences and when we talk about lower airway disease between the large, lower airways and the smaller, lower airways. Small or peripheral lower airway disease is often thought of as asthma. And it presents later in life. It's often acquired, produced by a trigger, accompanied by hypoxemia. There could be because of outflow obstruction, depressed diaphragm, there's a hyperinflation and has a good response to bronchodilators.

Large airway disease is tracheobronchomalacia. It presents earlier in life. It's often congenital. Can be accompanied by chronic congestion. There's usually no change in oxygen levels, no hypoxemia.

There can be palpable [INAUDIBLE], a normal diaphragmatic position, and no response to bronchodilators. These are clues that can help differentiate asthma from a larger, lower airway disease involving the trachea or bronchii.

So when we work up children with airyway abnormalities, it's important to do a good history and physical. And I'm not going to go through everything that you need to ask about. But a lot of it, it's on the slide. But things like a birth history, the age of symptom onset, associated feeding and voice problems, are important to ask about.

You want to observe the child breathing, phonating, and feeding, careful head and neck examination. Observe if there are retractions. How urgent is the situation? Is the patient stable? There's some utility in getting a chest x-ray or lateral neck x-ray sometimes, but it rarely gives a definitive airway diagnosis.

So really you have to think about when is endoscopy indicated. And endoscopy-- whether it be flexible endoscopy or rigid endoscopy-- is really indicated whenever the diagnosis of the cause of noisy breathing is in question. Or during emergencies because rigid endoscopy during emergencies can be used as a diagnostic, as well as a therapeutic, technique.

So when you see a patient in the office or in the emergency room, there needs to be an immediate assessment of the urgency of the situation. The patient is expectant. Are they tachypnic? Do they look in respiratory distress? Are they fatiguing?

Is the alar flaring? Is there use of accessory neck or chest muscles? Is there any evidence of air hunger? Do they look like they're starting to tire out? Sometimes when there's air hunger patients may hyper-extend their neck.

If they're unstable and you're really concerned, they need to be taken immediately to the operating room. If the child is stable, you can do a little bit more investigation. Carefully listen over the nose, mouth, neck, and chest. Look at the respiratory cycle. Is there any relationship with the stridor to the phase of respiration, because as we talked about a few slides ago, that can give us some clues to the cause of obstruction.

As I mentioned, supraglottic and glottic obstruction tend to lead to prolonged inspiration. And bronchial obstruction tend to lead to prolonged expiration. Interestingly, if you put the child in a prone position, some causes of stridor tend to get better. So if stridor's from the laryngomalacia, a large tongue, a short jaw, or intravascular compression in the trachea, children sometimes get better in a prone position. This isn't a gold standard and it can provide some extra clue to the cause of obstruction.

When there's respiratory distress with feeding, we have to think about a variety of different causes. Patients that are infants need to carefully coordinate their sucking, swallowing, and breathing. Sometimes when there's respiratory distress or difficulty breathing, they have difficulty with that coordination and can have swallowing problems. It can go both ways. Patients with swallowing problems can also have in concomitant airway problems.

So you can think about congenital nasal obstruction, tracheoesophageal fistula, vocal chord paralysis, laryngeal collapse, and reflux causes respiratory stress with feeding. If there's distress after a surgical procedure, of course, we think about could there be edema of the subglottic airway from the endotracheal tube. And there are different types of airway endoscopy. There's flexible endoscopy and rigid endoscopy. We'll talk a little bit more about

these two techniques, but they're important to know about, and they're often complementary.

The choice of which type of endoscopy to perform depends on the nature of the child's noisy breathing. If it's snoring or stertor or inspiratory stridor, often the diagnosis can be made from a flexible scope. If the patient's had intubation and you're worried about intubation injury, if the stridor's expiratory or biphasic, or if the patients are really complex-- a lot going on-- sometimes those patients benefit from a rigid endoscopy. I do want to emphasize that these two techniques can be complementary.

DAVID: So Jeff, this is a good time to stop. Take a little break. And so those of you that are watching and have questions, by all means, go ahead and submit them.

But I have a question for you, Jeff. So let's say that you're in a facility that's in a clinic or an office that's removed from a hospital. You just can't run up the stairs like we do at the VA. How do you make the decision-- whether to call 911 and get an ambulance, whether you need to just tell the parents to stick them in the car and take them on in? How do you decide that?

JEFF SIMONS: Well, Dave, that's a great question, David. I think that most of us have practices where we are seeing patients not in a hospital setting. We have some offices that are in the hospital and others that have a hospital nowhere close and we see patients and we don't have an emergency room close.

I think we have to use experience and judgment and carefully assess the children. If they look like they're in respiratory distress, if they're working really hard to breathe, if there's really significant retractions, I think those are reasons to consider having a patient be transferred to the emergency department. Other things you can think about are getting good vital signs, including pulisic symmetry disease, does the patient have any signs of hypoxemia.

And most of the patients that we see in the outpatient setting do not wind up having to go to the emergency room. They can continue their work up for noisy breathing in elective fashion. But every once in a while, and most of us have seen this happen, where we do see a patient that we're really concerned about.

They're worried that they're not going to be able to maintain their oxygen levels and they're going to tire out or they're going to become completely obstructive. And those are patients that we take immediately to the operating room. And so sometimes if you're in an outpatient setting, not at a hospital, those patients need to be sent via emergency medical services to the

hospital.

DAVID: Thanks.

JEFF SIMONS: Should we keep going? Again--

DAVID: Yeah, let's keep going. So feel free to send in questions and I'll interrupt Jeff as it seems appropriate.

JEFF SIMONS: I want to encourage you all to send in questions. You can also send in jokes. If they're good jokes, we'll read them to you. And we can share that with everybody.

So we can talk about diagnosis by site. The next few slides are going to list the differential diagnosis by site of obstruction. I'm not going to read the slides. They are on the website. You guys can access them later.

But we're going to go through this in a systematic way. And there is a lot of information here. We're not going to be able to cover quite all of it, but I want you to take home a general approach to children with noisy breathing and to be able identify the common causes of noisy breathing by anatomic site.

So we'll start with the nasal cavity-- pyriform aperture-- and in front we'll start with the anterior nasal cavity. Pyriform aperture stenosis is a congenital narrowing of the pyriform aperture. It tends to be narrowing in the front of the nose. It's often related to overgrowth in the nasal processes of the maxilla and interestingly, it can be associated with a single, central incisor. Patients are sometimes born with a single, central incisor.

And when you see that, they can also have accompanying holoprosencephaly and pituitary abnormalities. Patients that have--

DAVID: I see that holoprosencephaly all the time.

JEFF SIMONS: I suspect that you see this a lot at the VA, in the geriatric population.

DAVID: What is that?

JEFF SIMONS: Well I think what's important to note here is that patients that have a single, central incisor and that have nasal obstructure for pyriform aperture stenosis need more work up. They need an MRI of the brain and they need to probably see endocrinology to evaluate their pituitary

function. So symptoms of pyriform aperture stenosis include respiratory distress, cyanosis, and feeding difficulties. It can be diagnosed by endoscopy, which gives hints that there's a narrowed nasal vestibule.

And then CT imaging is the gold standard for diagnosis. And there's some CT criteria which are a little bit variable to define pyriform aperture stenosis. A narrowing less than 11 millimeters is the classic definition. But usually it's less than five when we really worry about those patients needing surgery.

There's different options for management of pyriform aperture stenosis. For mild cases, they can be observed. Intermediate cases sometimes benefit from the dilation of the nasal cavity with rigid dilators. And then more severe cases, can undergo other types of surgical correction-- formal surgical correction-- with a sublabial approach.

The picture there is an example of pyriform aperture stenosis. The arrows are a little bit off, I apologize. And on the right side of the screen is a sublabial approach to repair.

Also in the front of the nose we can see nasolacrimal duct cyst, also called dacryocystocele. This is a cystic dilation beneath the inferior turbinate. And it's usually caused by obstruction of the lacrimal duct valves, either the distal valves of Hasner or the proximal valves of Rosenmuller.

They can be unilateral or bilateral. But most of them, luckily, are unilateral. And the reason is that bilateral cases can be life-threatening because infants are preferential or obligate nasal breathers.

The treatment is excision of the cyst or marsupialization of the cyst and often accompanied by lacrimal duct stenting, which is done by a pediatric ophthalmologist. It's important to have a differential diagnosis for nasolacrimal duct cysts-- Other nasal masses-- congenital nasal masses-- like gliomas, dermoids and cephalocoeles, or even rhabdomyosarcoma. and because of that we usually like to get a preoperative CT scan before taking the patient to the operating room for marsupialization of the nasolacrimal duct cyst.

DAVID: Because if you find brain tissue, you know that's not what it was.

JEFF SIMONS: Then you know it wasn't a nasolacrimal duct cyst, absolutely. This is an example, on the lower left picture of the screen, is a picture of a nasolacrimal duct cyst. You can see it's located in the inferior meatus. And it's being marsupialized if you look in the right lower side of the

screen, opened with a sickle knife.

And there's more common causes of nasal obstruction that can lead to noisy breathing, as well as the inferior turbinate hypertrophy, which can be seen in all ages. We could see it in infants from neonatal rhinitis, which is often secondary to reflux, but sometimes it's idiopathic. You can see it in school-aged children from allergic rhinitis, and even younger from allergic rhinitis and you see--

DAVID: We see it in veterans.

JEFF SIMONS: In old people, also, don't you? And in veterans, absolutely. And then there's neonatal septal deviation, which occurs in approximately 1% of births, usually in higher incidence in vaginal births than C-sections. And It is most likely related to birth trauma, but there may also be some patients that develop in utero-- these neonatal septal deviation because of pressure of the nose, again something in utero.

It can cause life-threatening airway obstruction if it obstructs both sides of the nasal cavity. If it's just mild, patients can be observed. If there are severe symptoms, the patient be taken to the operating room within three days and the septum can be manipulated back onto the maxillary crest. Often, in order to get it to stay there, the neonatal nose has to be stented for a few weeks. This is an example of a neonatal septal deviation.

And then of course, there's sinonasal polyposis. Polyps are common I suspect in adults. You probably see them much more commonly in the adult population than in children.

DAVID: And we don't think of cystic fibrosis.

JEFF SIMONS: But in children we do still see them. And most of the times we see polyps in children, it is from cystic fibrosis, but not always. But the pediatric population, when you see someone with cystic fibrosis, they really do need to be worked up for cystic fibrosis.

And another 11% of cases are associated with asthma. As you know CF is a disorder of mucosal chloride secretion. It leads to impaired mucociliary clearance. And the patients tend to have chronic sinusitis, hyperplastic mucosal changes, and polyposis.

About half of children with sinonasal polyps-- About half of patients with cystic fibrosis have sinonasal polyps. And about 22% have undergone sinus surgery at some point and the average age is around 10. After they undergo surgery, or even before, they can also be

treated medically with topical and injected steroids and antibiotics. And it's important to tell the families that the reason to do surgery is to improve medical delivery-- topical medical delivery-- and improve patients' quality of life. But the surgery is often not curative and about 50% needed a second surgery within 24 months of the first surgery.

DAVID: So they still do sweat chloride tests? Diagnose--

JEFF SIMONS: Absolutely, so cystic fibrosis testing is part of neonatal's screening before patients leave the hospital when they're born, at least in the state of Pennsylvania, and I believe most states at this point. But there are still patients that could be not picked up that way. And so when we see patients in the pediatric population with polyps, we still do sweat chloride testing for CF.

There's also more advanced genetic testing that sometimes the pulmonologist will order as well. This is an example of polyps in a pediatric patient. You've got to think about intranasal foreign bodies, which we do see more commonly in children than adults, and most commonly in young, under age five, and developmentally delayed children.

And the most common symptom is unilateral [INAUDIBLE] nasal discharge, so foul-smelling smelling rhinorrhea. They tend to be located below the inferior turbinate or just anterior to the middle turbinate. And they can be there for a long time if not picked up.

The ones that are most dangerous are vegetable matter and button batteries. Button batteries cause caustic injury within an hour of being there and they have to be taken care of right away as an emergency because they can lead to significant mucosal injury and septal perforation.

DAVID: Now is that due to the acid leaking out of the button battery?

JEFF SIMONS: Well that's a whole other conversation. We would even probably need a whole other webinar in order to discuss all the details of button batteries, but that's one of the reasons. But most likely there's an electrolytic current that's created at the negative ion pull of the button battery that creates a local production of hydroxide radicals, which causes the caustic injury. It used to be thought that it was just from the substance leaking out of the batteries, but it's probably more from the electrolytic current and hydrolysis.

DAVID: So you make the strong base. But it matters.

JEFF SIMONS: That's controversial, but in the past couple of years, that's what researchers are thinking in terms of the mechanism for button battery injury. So generally anesthesia may be required for

removal of foreign bodies in some children. We try to take care of them in the office or in the emergency department when we can. This is an example of a nasal foreign body.

Then we'll go back towards the posterior aspect of the nose. The choanae is the part of the nose that connects the nasal cavity to the nasal pharynx. I want to emphasize that neonates are preferential nasal breathers for the first six to eight weeks of life. Because of that, if they have complete nasal obstruction, it can be life-threatening.

Choanal atresia is the failure of the choanae to open. And like I told you, the choanae is the part of the nasal cavity that separates the nasal cavity from the nasal pharynx posteriorly. Choanal atresia is present in about one in 5,000 births. 45% are bilateral and 55% are unilateral. The ones that have to be dealt with more urgently are the bilateral ones, again, because of the preferential nasal breathing in infants.

They can be membranous, bony, or both, but most of them are some mixture of membranous and bony. And there's different techniques for repair. In the old days they were done transpalataly. In fact, even in my training, in residency, and in fellowship, I did a few with the transpalatal approach. Most in the year 2016 are done endoscopic transnasal.

And then post-operative patients are given some steroid sprays to help prevent recurrence. They can be taken back to the operating room for removal of granulation tissue. And there's some discussion about whether or not they need to have stents in place. It used to be that everybody had stents in place, had [INAUDIBLE] repair, and there's now a trend towards to trying to avoid stenting when possible. That's still a little controversial.

Choanal atresia can be associated with some syndromes including, CHARGE. This is an example of a unilateral choanal atresia on the CT scan on the left, and it's being repaired on the right. And on the lower right side of this picture is a healed, repaired choanae after a choanal atresia repair.

And then there's common things we could think about, in terms of noisy breathing, things like adenoid hypertrophy. And they can be diagnosed with nasal endoscopy or it can be diagnosed with a lateral neck x-ray. And both those are successful techniques. And adenoid hypertrophy can lead to snoring, chronic nasal congestion, stertor, and airway obstruction.

Also, tonsillar hypertrophy is a common cause for snoring, stertor, obstructive sleep apnea in children. It's a common cause for noisy breathing-- something we need to always be thinking

about. And then there's midface hypoplasia. So these are some of the less common things that are more tertiary.

Midface hypoplasia can be seen in a variety of syndromes. It can lead to the collapse of the soft palate. It can also exacerbate obstruction at the level of the nasal pharynx. It can be associated with a whole variety of different syndromes. And we're not going to go through all the details of these syndromes, but there's syndromes like Crouzon syndrome, Apert's, Pfeiffer's, Treacher Collins, and Down syndrome.

Down syndrome can be associated with a variety of other airway abnormalities as well, including hypotonia, obstructive sleep apnea, and increased incidence of subglottis stenosis, which we'll talk about a little bit later in this presentation. And then there's Micrognathia. Micrognathia-- also related is retrognathia. Whether the jaw's small or protruded, it can lead to base of tongue collapse.

And that can be associated with Pierre Band Sequence. Pierre Band Sequence is a triad of micrognathia, glossoptosis, which is collapse of the tongue, and airway obstruction. It's also often accompanied by a cleft palate. So you can find micrognathia in a variety of syndromes-- Cornelia de Lange, Nager, Treacher Collins, and Stickler. But the one of the things we think about it is Pierre Band Sequence, which again, is a sequence not a syndrome, but can be associated with some syndromes such as Stickler syndrome.

Micrognathia, if it's mild, can be managed conservatively. Sometimes patients are dispositioned. Sometimes they may need some help with feeding for a little bit of time or sometimes the nasal trumpet. For more severe cases, they may need surgery, either tongue-lip adhesion, mandibular distraction, or in severe cases or when there's mulitlevel airway obstruction, a tracheotomy can be considered.

Mandibular distraction has become more and more popular when we're dealing with like micrognathia and tongue base collapse. And in that technique, Salter osteotomies are performed and the jaw is slowly distracted and then out for the bone to consolidate. It has the benefit of moving the jaw forward and also moving the tongue forward and actually the distraction part happens fairly-- I said slowly-- but actually the distraction part happens fairly rapidly. The consolidation is what takes time. The distractors are left in place while the bone grows and consolidated.

And then there's pharyngeal soft tissue obstruction, which you can see in trisomy 21, cerebral

palsy because of decreased [INAUDIBLE] muscle tone, and in certain mucopolysaccharidoses, like Hunter and Hurler syndrome. These are all things that we certainly see in a tertiary care pediatric otolaryngology practice and probably even in the general otolaryngology practice. Every once in a while people will come into your office with these types of problems.

In terms of the collapse of the epiglottis tongue base, there's a grading system, which one of my former colleagues here, Dr. Yellen, came up with, and it's widely used now to degrade the severity of epiglottic tongue base collapse. These are some examples of grade one, two, and three, three being the most severe with the tongue base [INAUDIBLE] against the posterior pharyngeal wall. The induce in the reflux can be associated with noisy breathing. So laryngopharyngeal reflux and the gastroesophageal reflux disease in children presents a little bit differently sometimes than with adults.

You don't always see heartburn and sour taste in the mouth and cough, sometimes you see airway symptoms. There can be stertor, cyanotic spells, failure to thrive, choking and gagging, and a lot of these symptoms are more common-- These types of reflux symptoms in children under age 2. A lot of times we'll empirically treat with positioning, sometimes thickening of feeding, and reflux medication. And sometimes a patient will be taken to the operating room for an endoscopy, or the gold standard diagnosis is pH metry, but it's not commonly used in the pediatric population. Often we start with empiric treatment to see if things will get better.

And then there's eosinophilic esophagitis which is defined as more than 15 eosinophils in a high powered field on esophageal biopsies, there's an eosinophilea of the esophageal mucosa and tend not to respond to proton pump inhibitors or other reflux treatment. It can be associated with a variety of swallowing and airway symptoms. It Can cause laryngeal inflammation. It can lead hoarseness, cough and stridor.

And the treatment for eosinophillic esophagitis is sometimes eliminating certain foods because it can be related to food allergies. An elemental diet can tend to be used or ingestion with fluticasone or budesonide, which is actually a swallowed corticosteroid.

And another interesting thing to mention about eosinophilic esophagitis is it can be associated with food impaction. So patients that have food impactions in the esophagus, particularly if they're recurrent food impactions, you always want to think about biopsying the esophagus to look for evidence of eosinophilic esophagitis is the cause for their underlying esophageal motility problem.

DAVID: OK before we go on, we do have a question, and this is a good time for a break because we're moving from the pharynx now to the larynx. So I have a question and the question has to do with, what are the criteria for mandibular distraction? So do you use specific measurements like a specific amount of retrognathia? Do you use measurements on [INAUDIBLE] or are there other findings that you use to trigger the decision to go with distraction?

JEFF SIMONS: I think that's a really good question. I think the first thing to think about is who is a potential candidate versus not a candidate. And there's some reasons some patients are not necessarily good candidates. I think some of the reasons that patients may not be good candidates is if they have central sleep apnea. If there's significant central sleep apnea accompanying obstructive sleep apnea, then taking care of the obstructive part may still not obviate the need for a tracheotomy.

So patients that have significant central sleep apnea, we sometimes think twice before they get distracted. Another thing is if there's multi-level airway obstruction, that even if you deal with that one level at this moment, it's still not going to deal with the several other levels of obstruction that are going to need a surgical airway at some point. That's another reason to think about waiting on distraction.

And the reason to distract early is to potentially prevent a tracheotomy.

If patients are not going to be able to benefit from that reason-- if they're going to need a tracheotomy for another reason-- then sometimes we'll wait. And some patients, by doing delayed distraction, the jaw grows and you can avoid the distraction. I don't personally do the distraction procedure, but the surgeons that do it usually get 3-D reconstructions of CT Scans with careful looking at the skeletal structure of the craniofacial bones and they determine the details of how to do the distraction. But as an otolaryngologist, I think the more important part is to determine whether distraction is going to address the most important causes of airway obstruction and prevent the need for a tracheotomy.

DAVID: So to take off on that last question, and that is, do you do DISE, drug-induced sleep endoscopy, in children, like is now being done in adults for sleep apnea? Is that a procedure used in children?

JEFF SIMONS: David, that's another great question. And drug-induced sleep endoscopy is absolutely a procedure that's used in children and we do it commonly. I would say we do it every week

several times. But we don't do it usually in the neonate with noisy breathing. They get endoscopy, but we're not doing it as a formal DISE, or drug-induced sleep endoscopy.

Typically, we're doing DISE in patients that have failed adenotonsillectomy. There was obstructive sleep apnea, despite having adenotonsillectomy or in certain patients with craniofacial abnormalities, craniofacial syndromes, Down syndrome, that their levels of airway obstruction leading to sleep apnea are multifactorial. It's a very good question. And we do do DISE in children.

Pediatric sleep surgery is definitely not as advanced as adult sleep surgery. There's a number of individuals in the country that are leaders in this field. And it's really blossoming a lot and there's becoming more and more literature out there about DISE in children.

DAVID: So we should go on.

JEFF SIMONS: Let's go. So we talked about the nose and the nasal pharynx and the oropharynx, the hyperpharynx. Let's talk about noisy breathing from the larynx at this point. And we can't have a talk about noisy breathing in children without talking about some of these common disorders.

The laryngomalacia is the number one congenital pediatric laryngeal disorder and the most common cause of stridor in infants. It's the most common cause. It's important for all otolaryngologists to know about the laryngomalacia.

It's caused by abnormal sensory motor laryngeal nerve integration and immaturity of superglottic cartilages and decreased laryngeal sensation. And that ultimately leads to a floppiness or collapse of the superglottic structures. Symptoms are inspiratoy stridor, which tends to be worse supine, worse doing feeds, usually worse with outset and crying. It's typically diagnosed with a wake flexible endoscopy in the office. It can be diagnosed in the OR, but the classic thing is an awake flexible endoscopy in an infant done in the office.

It's intimately associated with reflux. More than 90% of patients with laryngomalacia only require observation or reflux treatment and most resolve by 18 to 24 months. We can do anti-reflux strategies like thicken feeds, we treat with reflux medications, if we need to avoid feeding too close to bed time, avoiding feeding too much at a single feed. And then surgical treatment for the laryngomalacia is really only necessary in about 10% or less of patients.

And the indications for surgery, while they're evolving, the classic indications are apnea, sionosis, failing to thrive, severe dysphasia, feeding difficulty, life threatening events, and one

of the things that we do at the same time as the [INAUDIBLE] is look for other airway abnormalities. Some percentage of patients with laryngomalacia have to concominate-- we call them synchronous-- other airway abnormalities.

The second most common pediatric laryngeal disorder is vocal chord paralysis. And about half are unilateral and half are bilateral. In unilateral the common symptoms are hoarseness and aspiration, etiology, most common is cardiac surgery. Neck trauma is the second most common.

It can be managed with swallowing therapy, injection laryngoplastys. In older children we do thyroplasty. We try to avoid thyroplasty in young children because operating on the laryngeal framework may affect growth of larynx. In addition, the position of the vocal chord changes and the larynx grows, and so often patients that are children that undergo thyroplasty, have to undergo revision thryoplasty. And then more commonly, and now, it would be because of the difficulty with thyroplasty in the pediatric population, laryngeal reinnervation surgery is becoming more popular in children for unilateral vocal chord paralysis.

For bilateral vocal chord paralysis, the most common symptom is biphasic stridor The voice qualities, often, can be very good because there's good addiduction in the vocal folds. Classic etiology for bilateral vocal chord paralysis is central nervous system abnormalities, like chiari malformations. Many of these are idiopathic.

It's important to palpate-- or some way to evaluate-- the larynx to rule out posterior glottic stenosis, which is scarring in the posterior larynx, which can look a lot like bilateral vocal chord paralysis. A lot of times if patients are doing well, they can be observed. And often have spontaneous recovery within 12 to 18 months.

About half of patients with bilateral vocal chord paralysis do require a tracheotomoy. In general, those are patients that are having a lot of airway symptoms-- stridor, feeding difficulty, respiratory distress-- and there's other surgical options that can be performed down the line-- transverse cordotomy, needle [INAUDIBLE] arytenoidopexy, vocal chord lateralization, or even a postriograph laryn and tracheal reconstruction to make more room in the back of the glottis. This an example of a CO2 laser [INAUDIBLE] cordotomy and the posterior [INAUDIBLE] plate with the cartilage [INAUDIBLE] is the bottom picture on your screen. So those are two different surgical procedures proposed for bilateral vocal chord paralysis.

Subglottic stenosis is the third most common congenital pediatric laryngeal disorder after laryngomalacia and vocal chord paralysis, but it's the number one acquired pediatric laryngeal disorder. And it's the most common laryngeal disorder requiring a tracheotomy in children. The etiology is, by far, most commonly subglottic injury from the intratracheal tube. 95% of these are acquired and only 5% of them are congenital.

It often presents with stridor after endotracheal tube intubation, can present with recurring croup, there can be life-threatening respiratory stress, and some of the patients will come to us already with the trach because they've been intubated and trachs will prolong ventilation. And then we pick up the subglottic stenosis incidentally, when they're having the scope to look at the way their trach is sitting and to make sure there's no trach complications. Management can involve observation in mild cases. A tracheotomy in very severe cases.

For acute stenosis, endoscopic [INAUDIBLE] are often beneficial-- things like balloon dilation, division of webs, and open surgery for more mature, significant stenoses and that can involve laryngotracheal reconstruction or [INAUDIBLE] resection. This is an example of congenital subglottic stenosis and on the right side is acute intubation injury. This is balloon dilation of a acute intubation injury. And this is what it can look like on the right side of the screen, after balloon dilation.

And then this is another example of a more mature subglottic stenosis. This is a grade three subglottic stenosis, which is between 70% and 99%, so it's probably about an 85% subglottic stenosis. And this patient underwent a open [INAUDIBLE] reconstruction with anterior and posterior cartilage graphs. In the upper right side of the screen, you can see the anterior cartilage graph being put into place.

In terms of post-extubation trauma, the instances of acquired subglottic stenosis has been between 0% and 4% over the last few decades. It used to be more common than that. It's likely decreasing [INAUDIBLE] because of better intensive care unit care, treatment of reflux during intubation, trying to decrease periods of intubation, and going to a tracheotomy for patients that need prolonged intubation.

And for patients that do have to post-extubation trauma, the progression of the initial injury is variable. It's hard to predict who's going to develop mature stenosis and who's going to get better. In terms of late subglottic or tracheal stenosis, a soft and thin stenosis can be managed by endoscopic dilation sometimes. But with this pupil dilated, sometimes incisions will be made

with a laser followed by a balloon dilation. And the more firm, or long stenoses need open airway reconstruction.

Posterior glottic stenosis, as I mentioned, is scarring in the posterior glottis. It can be just a band between the vocal process of the retinoids, or you can have bilateral [INAUDIBLE] joint fixation or anything in between. And there's different treatment options for this as well, but it can contribute to poor abduction in the vocal folds and noisy breathing.

And then there's laryngeal webs, which is failure of embryologic reconalisation of the larynx. It's usually glottic, but it can extend to the subglottis. If it's mild, the symptoms can just be a breathy voice, but severe laryngeal webs lead the airway obstruction. They can have stridor obstruction, and may even need a trach.

Management is pretty complex in terms of who needs what type of treatment. Mild ones, in general, it can be incised and dilated. Sometimes there's application of mytomicin C. And in terms of open versus endoscopic approaches, that's a little bit outside of the scope of this talk. But I'm happy to discuss it if anyone has any questions about it.

And then there's laryngeal atresia. These are patients born with complete obstruction of the larynx. It's associated with CHAOS, which is Congenital High Airway Obstruction syndrome. And often because of the congenital high airway obstruction, there's intraurine polyhydramnios and fetal-- high drops. And that develops because the fetal pulmonary secretions are trapped in the lungs, they expand, and they compress and return to the heart.

These patients can only survive if they're delivered via an exit procedure. In an exit procedure, the airway is secured while the patient is still attached to the mother-- a c-section is performed-- and the airway's secured either via tracheotomy or intubation. And with CHAOS, it has to be a tracheotomy, relying on utero-placental circulation during the time of the surgical procedure. These are some pictures of us in the operating room doing an exit procedure. The ones on the right are large, surgical facial teratomas that are contributing to an airway obstruction.

We should talk a little bit about recurrent respiratory pathalomotosis. Again, I'm not going to read all the information on the slide. You can refer to it at any point. It's the most common neoplasm of the upper airway in children. Likely it comes from vertical transmission from an infected mother.

It's commonly associated with HPV type six and 11, as opposed to cervical cancer or pharyngeal cancer, which is more commonly associated with type 16 and 18. The prevalence is about four in 100,000 children. It can have symptoms of hoarseness or airway obstruction.

The first line of treatment is to remove the obstructing lesions and there's a variety of techniques to do that. We often start with a [INAUDIBLE], a pulse KTP laser, or CO2 laser can be done. And patients often need interative treatment, multiple treatments. Patients that need a certain number of treatments or if they're really having to go back to having to rely, there's adjustmen therapies we can consider.

And some of those are topical. Some of them are systemic. And the common ones used now are cidofovir, Avastin, interferon, which has become less popular now that there's better other options, and even indole-3-carbinole, which is a vitamin supplement found in cruciferous vegetables.

There's new HPV vaccines out there, which are now not so new. Gardasil is four valence vaccine, that's FDA approved between ages nine and 26. And hopefully we can make RRP less common as we move forward.

DAVID: Jeff, is there any data that there's been any change in the incidence of RRP since the advent of these vaccines or it too early to know?

JEFF SIMONS: It's a great question. And several questions related to this come up. The answer is it's too early to know at this point. But anecdotally, a lot of people think that we're seeing less incidents at this point.

Another good question that people don't know the answer to, is what's the benefit to taking children before that age of nine that already have RRP and giving them the vaccine? Is there any benefit to vaccinating children under age nine that already have RRP? And some of the benefits are that it may prevent them from developing other types of the virus as they go forward. But the question is, does the vaccine change the immune modulating function that would help improve the patient's current disease status? And that's something that's being investigated both in the US and in Europe.

And there's subglottic hemangiomas, which are the most common neoplasm of the infant airway-- for the infant airway, that is. They grow rapidly in infancy, then slowly regress. They present with progressive biphasic stridor. If they're not addressed, they have a high mortality

rate, but that's not common anymore. They're almost always treated at this point.

About half the patients that have a subglottic hemangioma may also have cumantaneous hemangioma, which is usually located on the neck or face. The diagnosis is usually made by endoscopy. You don't need a biopsy it. It can usually be visualized.

And if you're concerned about there being transtracheal extension into the neck or intrathoracic region, then imaging is necessary, otherwise probably not. Not all patients with subglottic hemangiomas need to have MRIs. Management is individualized. There's a variety of treatments. Very mild ones can be observed.

If they're severe, it can be obstructing. A tracheotomy is an option. There can be systemic or local steroids, open excision with laryntracheal reconstruction. And the most exciting thing that's come out over the past few years, is propanolol, which is a beta blocker, which has been very successful in treating many hemangiomas in infants. And our current protocols for treating subglottic hemangiomas involve the use of steroids and propanolol

And then there's tracheobronchomalacia, which is the floppiness of the tracheal and bronchial cartilage. The airway collapses more in expiration, tend to have stridor, and respiratory distress. Again, it's more commonly excretory noisy breathing with trancheobronchomalacia.

Primary tracheobronchomalacia is an inherent cartilage weakness leading to collapse. It usually gets better as patients get older. The cartilage matures and becomes stiffer. It can be managed by observation, sometimes inhaled ipratropium can be beneficial for mild to moderate cases. And if it's more severe, treatment with positive pressure therapy is a possibility.

Tracheobronchomalacia can also come in the extrinsic compression form, which either be from a extratracheal mass or vascular compression, which is often from innominate artery. Management, if it's mild, can be observation. If it's severe and it's from vascular compression, you can do an aortopexy, which can help lift innominate artery off the trachea and relieve the compression. And also you can have tracheobronchomalacia acquired from a tracheotomy. And the other term we use for that is superstomacartilaginous collapse.

Management of that can be endoscopic versus open repair. This is an example of primary tracheobronchomalacias. On the upper part of the screen is extrensic vascular compression. And in the lower part of the screen is the suprastomacartilaginous collapse, which is very

common in children with a tracheotomy. How often you see that in adults with a tracheotomy-suprastromacartilaginous collapse?

DAVID: It does occur. We do see that.

JEFF SIMONS: It's interesting because of the relative immaturity of cartilage in children and the smaller airway, there's some complications of trachs in children that we see with a much higher rate than in adults. And so we do more interval surveillance of the trach sites in the airways in children with trachs, compared to what's done in a common adult airway or a tracheotomy practice.

Tracheal rings are supposed to be C-shaped and have a [INAUDIBLE] aspect. You can see complete rings and they often associate with tracheal stenosis. They can accompany vascular abnormalities in the chest. And can be seen with certain syndromes, such as Down syndrome. You often see them with trachesophogeal fistulas.

Complete tracheostenosis or complete tracheal rings, it can cause a long segment stenosis. It can be significant. And if it's mild, the treatment is observation. If it's a short segment, relatively, the [INAUDIBLE] can be resected with an endoastamosis.

Other options for longer segments, the most common, exciting thing now is the slide tracheoplasty, which is particularly good for funnel-shaped stenosis or a long segment stenosis. On the right side of the lower picture, there's a nice example of complete tracheal rings.

Interestingly, that patient has had asymptomatic complete tracheal rings. They were picked up when I took the patient to the operating room to remove a foreign body-- an airway foreign body-- that was aspirated. It was a boy that was chewing on his pen cap. He aspirated his pen cap.

We went to take the pen cap out and then incidentally found the complete tracheal rings. And this is an example of a patient while on cardiopulmonary bypass, undergoing a slight tracheoplasty. We do this procedure together with the cardiothorastic surgeons. We often work as a team. And the patients are on bypass for the slide tracheoplasty procedure.

Foreign body aspiration often has to be considered in the differential diagnosis of noisy breathing in infants and children. It most commonly occurs in the one- to three-year-old population. Almost always under age 5, but not always. There's also a group of adolescents

that chew on their school supplies and aspirate them.

It accounts for about 7% of lethal accidents int the one- to three-year-old population. The most common site is the [INAUDIBLE]. There could be acute symptoms like coughing and choking. And then longer term symptoms of the foreign bodies aren't diagnosed, like a persistent cough, wheezing, pneumonia, and [INAUDIBLE] develop as inflammation and granulations to develop.

If there's a witness aspiration, It's the most predictive of the diagnosis, but these are often not witnessed. So in the absence of a witness event, you need to have a high index suspicion. The findings on a chest x-ray, you can see air-trapping, adolectasis, but in terms of seeing an actual foreign body or a radio-opaque foreign body, it's really only 15% or so of patients. Most of the aspirated material in kids are food-- nuts, seeds, corn kernels-- and most of them are radio-opaque. And the treatment, both diagnostic and therapeutic, with rigid ronbracheal removal.

I want to spend the last few minutes here talking about some infectious airway disorders. We alluded to a little bit to croup earlier in the talk. Croup is laryngotracheal bronchitis. It tends to peak in age between one and two years.

And that's because of the small size of the airway. Remember the neonatal airway is really four to five millimeters in diameter. It's usually viral and the most common virus is parainfluenza. But influenza in RSB can also cause croup.

As a clinical diagnosis, you don't want to take patients with acute croup to the OR, if possible, because if you instrument an already andaminas-inflamed airway, you can make it worse and patients can wind up intubated in the ICU for a long time while they're recovering. It can be accompanied by a low-grade fever, a barking cough. It's usually biphasic stridor. Usually most patients with croup can be treated as an outpatient with humidified air, oral steroids, and in more severe cases, sometimes you need to go the emergency room, and rarely, to be admitted. And the patients, when they go to the emergency room will be treatted with racemic epinephrine and intravenous corticosteroids.

So when patients have recurring croup, they're often referred to us as pediatric otolaryngologists for further work up. And really the things we're looking for is, is there underlying other abnormalities we need to pick up on. And they're things like subglottic

stenosis, laryngomalacia, subglottic webs. All those things occur in about 25% of patients with recurrent croup and reflux is actually quite common in patients with recurrent croup, probably more than 50% of patients.

Then there's bacterial tracheitis, which was initially thought to be rare, but probably is more common than we thought, and is probably somewhere on the continuum with viral croup actually. The average age is a little bit older, between four and six years, most commonly from staph areus. The symptoms are high fever and stridor. You see the thick, membranous tracheal secretions that can obstruct the airway.

Treatment for severe bacterial tracheitis is a prompt trip to the operating room for an endoscopy and removal of those obstructing secretions, in addition to treatment with intravenous antibiotics. So in general, it seems like the severity is decreasing and this is being more commonly diagnosed at this point. Do you see it in the adult population--

DAVID: Believe it or not, I had a case last week. Grute proteus mirabilis. White count was 21,000. And he had a trach, but he had severe bacterial tracheitis.

JEFF SIMONS: Patients with croup that don't seem to be following the normal course of convalescence, you have to think about could the be something more than croup? And some of these patients, that would be a reason they would need to go to the operating room to look. And bacterial tracheitis needs to be in the differential diagnosis.

And then of course, there's acute epiglottitis, which I've seen two or three times in the 10 years I've been here as an attending. It's pretty rare now. There's rapid onset inflammation of the supraglottic larynx. It's a true airway emergency. The management a right right to the operating room, attempted intubation, and you're said to of obtain an airway whichever way possible, including a surgical airway if need be.

It used to be most commonly caused by H-flu Type B. I suspect when you were a resident, you saw this much more common than we do now. After [INAUDIBLE] vaccine, which came out in 1987, we still do see it and we see it most commonly in the unimmunized population. So the few cases I'm thinking about were all in Amish children that were not immunized.

And we can also see more mild supraglottitis, which is caused by other bacteria, like staph aureus. This is an example of the classic thumbprint sign you can see the left side of the screen or on a lateral neck film and a [INAUDIBLE] epiglottis and epiglottitus. You notice we

took the picture both before and after intubation. I felt a little better taking the one after intubation though.

So in conclusion, there's a variety of causes of noisy breathing in infants. It's important to identify the type of noisy breathing. The diagnosis is based on history and physical exam, as well as endoscopy, and that's both flexible and rigid endoscopy, and those can be complementary techniques.

The management options are based on the severity of symptoms, the diagnosis, and the general health of the child. And we're still searching. There's a lot of things we don't know. We're still searching for optimal treatment for many pediatric upper airway disorders.

We covered a lot of information in the past hour. Please free to send us any other questions if you have them. And you can also email me if you think of other things in the future at jeffrey.simons@chb.edu. And the slides will be available on the website for you to refer to in the future.

So Jeff, thank you very much. We have a few more minutes if anyone has any additional questions. Watching these slides and listening to Jeff, it brings back memories, not all of which were happy memories. And I think almost everybody who has been in practice for a number of years, of they took care of children, has had at least a couple of stories that they remember vividly.

DAVID: My favorite story about noisy breathing was a patient of [INAUDIBLE] who came in with a whistle. And it was an inspiratoy and expiratoy whistle. And it turned out that the patient had aspirated a Barbie shoe. It was a shoe from Barbie and it acted like a whistle. And when you listened to this child's chest, you could hear the whistle in and out. That was my favorite noisy breathing of all time.

JEFF SIMONS: It's funny that you mention a whistle because I take care of a lot of children with foreign body ingestions and aspiration. And I, just a few months ago, had a patient that came in, after she was playing with her dog and the dog had a squeaky dog toy in his mouth. And this pre-teen decided that she also wanted to put the dog toy in her mouth, so they both had it in their teeth and they were going back and forth.

And this dog toy had a squeaker and it was a little piece of plastic with a little flap that moved back and forth to make a sound. And this girl had that squeaker pop off of the dog toy while

she had it in her teeth, and she aspirated it. As she presented to the emergency room with a squeaky sound. Every time she breathed in, she squeaked. And that's what her mom said when she came in.

And when you listened to her, she actually squeaked, you didn't need a stethoscope. You could hear he squeaking. And sure enough, she had it in her right mainstem bronchis-- the little piece of the dog toy-- and every time she breathed in, the flap went back and forth until we took it.

DAVID: You got me beat, Jeff. That one beats the Barbie shoe. So I think we got about three or four minutes. And feel free to send us any more comments or questions.

You should know that at the ABEA meeting every year at COSM, they always have a Foreign Body of the Year contest. I remember one was a patient who had inhaled a cap from a tire valve and they had inhaled the cap and it had landed and impacted with the thread side up. So the otolaryngologist whose patient it was, had his father braze a valve stem piece on a instrument, which she put down, screwed into the cap, and pulled the whole thing out. He won the prize that year.

JEFF SIMONS: That's a good story.

DAVID: Oh we have another question or story. Let's see what we have here. When a foreign body does not need to be removed. So say if somebody swallows something or inhales something that's going to dissolve, do you ever use inhaled humidification? Are there instances when you don't have to remove it?

JEFF SIMONS: It's a really good question. So I suspect that there might be some rare instances. The only types of incidences will be certain food particles, but the thing is, most food particles that are aspirated tend to be things that have a lot of oils in them-- nuts and seeds. And if they're not removed-- even if there are portions-- not the entire thing-- they can cause a lot of local, chemical pneumenitis, or inflammation in the lungs.

And the patient can get sick. They're not so much having problems with airway obstruction, but they can develop pneumonia or pneumenitis down the line. So in general, even if food particles are small, we tend to try to remove them. There might be another question even.

DAVID: Nope, another question. Here we go. Just a comment. So thank you for your comments and questions. Well, with that, I think we should close. I think our time is up and they're about to

turn the computer off.

JEFF SIMONS: So thank you all for spending the evening with us. It was fun. Again, feel free to contact me if you have any questions and we look forward to seeing you sometime soon.