



Episode 14:
Pheos & Paras
Transcript

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Welcome to NET Wise. This is a podcast for neuroendocrine cancer patients and caregivers that presents expert information and patient perspectives. My name is Melissa Phillips, from the Neuroendocrine Tumor Research Foundation.

In the previous episode of this series, which was all about the relationship between NETs and genetics, we heard a bit about tumors known as pheochromocytomas and paragangliomas, often referred to as "pheos and paras". These are particularly rare NETs, affecting only maybe one in every 3,000 people, but are very strongly connected to some of the genetic conditions we heard about in that episode, including M.E.N. 1 and 2, Von Hippel Lindau Syndrome, and the SDH gene mutations. Here's Bonnie Bennett, Nurse Coordinator for the Neuroendocrine Tumor



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program at the University of Pennsylvania, and Dr. Lauren Fishbein, an Endocrinologist at UCHealth and the University of Colorado School of Medicine:

Bennett: *"Of all of the tumor types there are for cancer, Pheochromocytoma and paraganglioma have the highest association with genetic syndromes, genetic causes of any other tumor type."*

Fishbein: *"Up to 40% of patients who have one of these tumors has a genetic predisposition"*

Bennett: *"And we know of right now, some 17 different genetic syndromes that can be associated."*

Pheos and paras are complex tumors that are quite different from other kinds of NETs, and so today we're going to dedicate an entire episode to their unique biology, symptoms, and treatment.

To understand Pheos and Paras we have to start by talking a bit about your nervous system. At the center of this system are the brain and spinal cord, and then extending out from these are a complex web of nerves, which carry electrical messages from the brain out to every part of the body, and glands, which send out chemical messages that are understood by all of your organs and tissues. Your neuroendocrine cells act as the translators between these two kinds of messaging.

Generally speaking, this system is divided into two parts - the "somatic" nervous system, which controls things you do consciously, like walking, talking, and eating; and the "autonomic" nervous system, which controls things that happen without thinking about them, like breathing and your heartbeat.

A subset of the autonomic nervous system controls the things our body does automatically when we are hurt, stressed, surprised, or afraid. These "fight or flight" responses are caused by release of catecholamines. These are hormones such as epinephrine, also known as adrenaline, norepinephrine, and dopamine. Here's Samantha Greenberg, a Genetic Counselor at the Huntsman Cancer Institute in Salt Lake City, Utah:

Greenberg: *"So that system that kind of makes you feel like you're having an adrenaline rush or the system that makes you calm down from that adrenaline rush. And a part of that system is those adrenal glands."*

Fishbein: *"Pheochromocytoma and paraganglioma, these are a rare type of neuroendocrine tumor that form from the autonomic nervous system. If they're inside the adrenal gland, it's called a pheochromocytoma. If they form in one of the nerve bundles outside from the skull base, all the way to the pelvis, those are called paraganglioma. So paraganglioma and pheochromocytoma look exactly the same under the microscope - can't really tell them apart. So they're just named differently based on whether they're in the adrenal gland or outside the adrenal gland."*

There are a number of factors that make pheos and paras different from other kinds of NETs. The first is that they are very difficult to accurately grade. With other kinds of NETs there are physical characteristics that a pathologist can use to determine how likely the tumor is to metastasize, but with pheos and paras that just isn't the case. Here's Dr. Joseph Dillon, an Endocrinologist at the University of Iowa:

Dillon: *"The grading system, which is what the pathologist sees, is quite different from the grading system for neuroendocrine or carcinoid tumors of the pancreas or small bowel. I don't think the grading system is really as well worked out in pheochromocytomas and paragangliomas. You cannot tell when you remove a paraganglioma or a pheochromocytoma whether it's going to eventually spread or not, so you can't actually tell based on the pathology whether it's truly benign or not."*

This is further complicated by the fact that these tumors, particularly paragangliomas, have a tendency to develop multifocal primaries, so it's not always easy to determine whether multiple paras are one primary tumor and its metastases, or multiple primaries that developed independently of each other.

Dillon: *"Yes, you can have a primary paraganglioma that then metastasizes to somewhere else in the body, but you can also have the issue of multiple paragangliomas occurring either at the same time, or are at different*

times over, over their lifetime. It's sometimes very difficult with a paraganglioma when you see a scan to say this tumor here is a metastatic tumor versus this is multiple primary paragangliomas."

Despite the fact that accurately grading these tumors can be difficult, some do behave as high grade and others as low-grade tumors. Particularly among paragangliomas, some are very aggressive and need immediate treatment, and some are so indolent as to require no treatment except observation for many years.

Dillon: *"Some paragangliomas, depending on where they are particularly in the head and neck area, can be so slow growing and not causing any problem that a wait and see approach is taken. So these tumors are frequently near nerves and big blood vessels. Surgery can be difficult if it's not done in a true area of expertise. So the cure may be worse than the disease itself"*

Other paras and almost all pheos absolutely require treatment, though. One of the main reasons for this is their tendency to cause serious hormonal symptoms.

All pheos and a good percentage of paras produce excesses of those "fight or flight" hormones such as adrenaline. Here's Dr.



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Nancy Sharma, a Medical Oncologist at Swedish Cancer Institute in Seattle, Washington:

Sharma: *"Even if people are not symptomatic, based on the amount of catecholamines being produced, they still are functional. They will still have catecholamine release."*

Fishbein: *"Adrenaline, which is the hormone that we use when we get scared. It makes our breathing go more rapidly, our eyes get wide, we start to get sweaty, we might get shakier, tremulous. That allows us to run away from whatever is scaring us."*

Overproducing these hormones can cause a wide range of symptoms, both physical and emotional. The first is high blood pressure.

Greenberg: *"What's unique to these tumors is that people have persistently high blood pressure, even on medications, even in other settings."*

Dillon: *"Blood pressure that's difficult to control. So people who get diagnosed with blood pressure perhaps at a younger age - they get on one agent, two agents, three agents, four agents for their blood pressure and their blood pressure is still poorly controlled. That can be driven by adrenaline, a continuous excess of adrenaline."*



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This is interesting because it's the opposite of the effect of some more common NET-related syndromes.

Bennett: *"Carcinoid Syndrome causes the blood pressure to drop. The adrenaline causes the blood pressure to go up."*

This excess of adrenaline can also cause a whole host of mental and emotional symptoms that can be very difficult to manage.

Fishbein: *"Think about how you feel when someone comes up behind you and scares the bejesus out of you - you feel it, you feel kind of a surge in your body, right? Sort of energy coming up. Patients with pheochromocytoma can have levels of adrenaline that can be two times the upper limit of normal to fifty times the upper limit of normal. So there can be really high levels of adrenaline that don't go away. It can be very mentally challenging for patients."*

Sometimes this increased level of adrenaline is relatively steady, causing a persistent feeling of stress and anxiety, and sometimes it surges periodically, causing episodes that can feel a lot like panic attacks.

Dillon: *"So sudden onset of heart racing, sweating, pounding headache, pounding chest (which we call palpitations), shakiness, and indeed a sense of true anxiety lasting anywhere from five minutes to fifteen to thirty minutes. Stress can then cause an extra outpouring of adrenaline, causing an episode like this. So it can*

either be spontaneous or being associated with some precipitating activity."

Bennett: *"A lot of the patients will be misdiagnosed for many years with anxiety disorders, different psychological... maybe even suicidal, we've had patients who were suicidal because nobody can figure out what's wrong with them."*

Some people experience these symptoms not as anxiety, but as anger.

Dillon: *"It can certainly cause people to have to have a sense of rage and lash out. There are people who have ended up in court or in psychiatric facilities and eventually get diagnosed as a person with a pheochromocytoma."*

Fishbein: *"Especially because it is rare, and so it's not always thought of as the first diagnosis. If someone presents with anxiety, rage, or anger, we think about depression and anxiety as the more common diagnoses, and those are going to be the more common diagnoses. But we have to consider that if it's associated, maybe, with high blood pressure, if it's associated with an already known predisposition syndrome, like VHL or MEN2, then we have to think about pheochromocytoma as well."*

Soto: *"So my name is Eli Soto. I'm currently coming to you from Fort Collins, Colorado. I'm 43 years old. So, I was*

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diagnosed with a stage three metastatic pheochromocytoma. It was on my right adrenal, and it metastasized to the retroperitoneal cavity, to the lymph nodes. And I am coming up, almost... this summer will be two years of remission.

You know, the Pheo just kind of sneaks up on you. You know, this is a type of disease that is insidious in every part of your life, but it's like a frog boiling in water. That's the best way I can put it. You know, it happens so slowly over time that you don't know you're in any real danger until it's a big, giant problem inside you.

It was one day where I was building a closet and I was going back and forth from upstairs to my garage. And I was leaning down trying to get my miter saw off the floor. I pressed my knees into my chest and I strained trying to get it up off the ground. And in that moment... I don't know if you ever see the movies where somebody gets, like, injected with adrenaline and you hear the 'boom, boom, boom, boom...' you know, the heartbeat and everything? Well, that's what I could hear inside my body. I could, it felt like a rising tide. It felt like blood pressure or any kind of pressure was just rising from my chest to my neck. And then all of a sudden there was a huge explosion in the back of my brain.

Now, one of the things that I hate about the literature right now is they refer to one of the symptoms as severe headaches. That's like getting your foot cut off and saying 'my foot hurts.' It doesn't do it an accurate description.

For the listeners out there, I think the best way to describe one of these events is if you've ever seen the movie Alien with Sigourney Weaver - do you remember how the little alien pops out of the chest cavity? That's what it feels like in the back of your head. It feels like something is trying to get out of the back of your head. So I remember grabbing the back of my head, falling on the ground screaming. That was the most intense pain I have ever felt in my entire life.

And as fast as it comes on, this "boom, boom, boom, boom" adrenaline rush and this explosion, this feeling like you're about to die, you can't speak to anybody during that time. It's so painful. It goes away almost as quickly as it comes on, as if it never happened. You can get up and just walk it off, as if that moment never happened. It's the weirdest thing.

Then that week, it just started to get worse - the frequency of it just started to get more and more and more until one night, it just happened when I was just laying in bed, just not doing anything. And I told my wife at the time, I was just like, 'Listen, something's not right. I got to go to the hospital'. I made it to the hospital. I remember holding on to the front desk. And that's when I had another event, just a huge explosion. I fell backwards. I just remember it being back in the ER and they were cutting my clothes off, and they did every test possible that night.

So when I went back to go speak with the neurologist after the blood and the urine, he said, 'Eli', he said, 'the amount of catecholamines you have in your system today is the equivalent of doing crystal meth every day of your life'. So you can imagine what I must have been like to be around.

And I look back and, you know, there's a lot of things that I'm not proud of and just the interactions with work colleagues, personal family issues and everything like that. I didn't have a lot of patience. I was constantly anxious. Like, you know, I was nervous about the world around me. Edgy, agitated. Towards the last three years, I only slept two hours a day. You know, that in itself makes the mind go a little bit, kind of crazy, you know? And it's significant how much it affects your personal life and the relationships around you."

After a long period of misdiagnosis, Eli's pheo was finally removed surgically. Since then, he's not needed other treatment, and the hormonal changes to his mood and personality have disappeared.

"Everybody that knows me that's still in my life from before and after tells me how different I am right now. They're like, 'We all notice it'. Like, 'You're, you're not

the same man you used to be at all, and much more pleasant to be around.

I feel like I feel in the moment, in the present, I feel fine now. There's always that fear in the back of my head that's like, you know, anytime I have a bad day or, you know, I get agitated with somebody, that's my first go-to thought like, 'Oh my gosh, is this coming back? Did they miss something?' If it does come back, knowing what I know now, I feel like I'm at the forefront of all the access to all the research that's been done - just the level of care that I would need to get to get this out of me - that even if it does come back, I know I'll be in good hands."

Because Pheos are often connected to genetic conditions that run in families, it's not uncommon for patients to suspect that strange mood shifts they noticed in their parents or grandparents might have been the result of a pheochromocytoma that went undiagnosed. Here's Doug Karle, who told us about his experiences with having Von Hippel Lindau Syndrome, a hereditary condition that can cause Pheos:

Karle: *"So my father, he never obviously knew what was going on, but he had a complete personality change at about age 40. And it was a... there's a thing called 'Pheo rage', which is you have all this adrenaline pumping through your body. My mom and I talked about it not too long ago. It was a prominent shift from like the greatest kind of coolest dad to like just this angry person. And he didn't have a*

lot to be angry about, I mean, he was successful in his professional life and I think we were good kids and but, ah, no, it was a complete shift. In hindsight, it becomes more clear."

When it comes to treating Pheos and Paras, the best first option is usually surgery.

Dillon: *"As a general rule, the only curative treatment for pheochromocytoma or paraganglioma is a surgical removal of the tumor."*

Fishbein: *"And so once we make the diagnosis, we find the tumor on imaging and make the hormonal diagnosis, then surgery is the mainstay of therapy"*

When pheos or paras are functional and causing hormonal symptoms, there are steps that have to be taken to make sure surgery goes well. This is because adrenaline spikes are caused by stress, and surgery is about the most physically stressful thing you can do to a body. So before a patient with a pheo or functional para is operated on, they have to be treated with special medications called "alpha blockers" that mitigate those adrenaline rushes.

Fishbein: *"Anesthesia, the surgery itself, cutting into the skin, these are physical stressors where a normal body response is to increase adrenaline. And so, if you already*

have a tumor that has high adrenaline, and now it's going to go even higher, we worry about a hypertensive crisis where the blood pressure gets so high it could lead to a heart attack or stroke. And so that's the reason to give this alpha blocker medicine before any surgery so that we can block those receptors as best as possible. And then during the surgery, the anesthesiologist will use similar alpha blocker medicines through the IV to help control that as well, because we don't want any adverse outcomes from the high adrenaline level during surgery."

Bennett: *"They will need to be on that medication usually for about two weeks before they have surgery."*

This is something that has to be kept in mind for pheo or functional para patients who are undergoing any kind of medical procedure that may cause physical or mental stress.

Bennett: *"Any procedures, like if they go to the dentist, they may need blocking because they have this low level of catecholamine release from their tumors, and we don't want them to have a problem when they go for a procedure. So, if they're going to have a colonoscopy, any type of procedure, they need surgical blocking."*

Dillon: *"One of the problems with diagnosing a paraganglioma and pheochromocytoma is that one of the things that can cause a pheochromocytoma to actually*

produce a hormonal surge is sticking a needle in it for a biopsy”

Bennett: *“If someone is in a car accident and they go for imaging and the imaging shows something on the adrenal gland, that should never be biopsied, because if it's a Pheo and your blood pressure shoots up, you would be at risk for having a heart attack or stroke. So radiologists should know that if there's something worrisome there on the adrenal gland or extra adrenal in a location where paragangliomas are known to be found, they should not be biopsied”.*

Once this preparatory work is done, the next question is what kind of surgery to perform. For Pheos, there are two primary options: total resection, where the entire adrenal gland is removed, or partial resection, where some of it is left intact. The choice largely has to do with whether your care team thinks it's likely that the cancer will reoccur.

You have two adrenal glands, one above each kidney, and if one remains in place, the body can function normally. If both end up needing to be removed, the patient will need hormone replacement therapy to make up for what their adrenal glands are no longer doing. So, if the doctors think it's likely that the surgery will be curative, they might recommend a total resection of the adrenal gland that has the pheo growing in it. If they see evidence that a second pheo might grow on the other side - because, for instance, the patient has a genetic condition that

increases the risk of further pheos developing - they might opt for a partial resection. Here's Dr. Nancy Perrier, Head of the section for surgical endocrinology at MD Anderson Cancer Center in Houston, Texas:

Perrier: *"We're going to want to start planning that operation to resect the tumor, but to do so in a way that leaves the least amount of morbidity in the long term, so we're always taking those risk/benefit ratios."*

The adrenal gland is like a boiled egg, a little boiled egg that sits right on top of the kidney, and a pheo is a tumor that arises in the yolk of the boiled egg. But the adrenal gland has a function for the white and a function for the yellow. So if we're trying to remove the tumor and let's say that was, you know, a cherry or a plum that was growing out of the yolk, our goal would be to remove the yolk and that plum, but to leave a little piece of the white boiled part of the egg and just the white, so that that patient would then have the hormone and would not need hormone replacement for the cortisol, which is made from the boiled white of the egg."

It might sound counterintuitive, but a total adrenal resection is a quicker and simpler surgery than a partial resection.

Perrier: *"It's easier just to take out the whole boiled egg, right? It's intact. It has good margins, and it doesn't fracture. If you have to start getting into carving*

the tissue and making sure that it doesn't fracture and whatnot... but if you're just gonna scoop and just take the boiled egg out, that can be quite easy. And so for a patient, when we're not worried and we know that that other gland is going to be fine, that can be a simple, easy operation.

A paraganglioma doesn't arise in the adrenals, so there is no role of saving any portion of that. The decision making has different branches, but it's not partial versus total resections, always total resection"

In a large percentage of Pheo/Para patients, one successful surgery may be all that is needed. That isn't always the case, though. As we've heard, people who have one of the genetic syndromes that's associated with these tumors are particularly prone to recurrence and need to be screened regularly to make sure new tumors haven't popped up.

Greenberg: *"When somebody has a paraganglioma or a pheochromocytoma identified, they need genetic testing to know if there is some kind of hereditary cause for that mutation, which as we've talked about can impact both treatment and family members. Once somebody has that mutation identified their family members also need genetic testing, which is typically a saliva or a blood sample to identify if they have that same genetic change that's previously been found in other relatives. After that, if*

somebody tests positive for a mutation, regardless of whether or not they have a tumor history, they require screening. And that screening is what we would consider to be high risk screening, which means you have an increased risk for these tumors, and we need to do imaging and blood work, or a urine sample, to identify whether or not you might have these tumors. And the goal of that screening is to identify those tumors early so that we never get to the point where patients have signs or symptoms of these tumors, but rather that we're able to identify them resect them, or monitor them without impacting the patient's quality of life."

And even in patients without a genetic syndrome, recurrence can happen, and sometimes many years later, so an annual screening is still recommended.

Fishbein : *"For most people with pheo/para, surgery is curative. We still recommend that everyone gets followed for life if they've had a pheo/para at least once a year, to just make sure there's no recurrence or another primary multifocal tumor."*

In cases where the tumors do recur, further surgeries can often be the best option.

Perrier: *"Yes, (it's) not uncommon that repeat surgery is a part of the lifelong management of these patients."*



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Sometimes, though, surgery isn't an option, or isn't sufficient to manage the disease.

Sharma: *"Of course surgery is the cure, but we know that up to 10-15% of Pheos and more of Paras - up to 30% of Paras - can be malignant, and they can come back, sometimes several years down the lane. And a lot of time, if they come back, and they are metastatic and local therapies are not an option in that situation, then we do get involved for systemic therapies."*

Nuclear medicine treatments are offering hope to patients with pheochromocytomas and paragangliomas that have spread and can't be removed surgically. Here's Dr. Erik Mittra, Chief of Nuclear Medicine and Molecular Imaging at Oregon Health and Science University in Portland:

Mittra: *"For patients who are inoperable for a variety of different reasons, whether the location of the lesion, or because it's wide-spread metastatic disease, either of those cases, then this becomes the next line of therapy."*

Nuclear medicine is available for Pheo/Para patients in two forms. The first one is PRRT or "Peptide Receptor Radionuclide Therapy", using a medication known under the brand name "Lutathera".

Dillon: *"These tumors very frequently - 80, 85% of them - will have somatostatin receptors. So PRRT, intravenous radioactive somatostatin, will stick to the tumor, bring the radioactivity into the tumor."*

The other option is one that was specifically developed and approved for use with pheos and paras and other adrenal tumors, such as neuroblastomas. It's called MIBG therapy, which uses a medication called "Azedra".

Mittra: *"So MIBG stands for metaiodobenzylguanidine. And that's the molecule, which now has the brand name Azedra, and that's attached to the isotope Iodine 131, which provides the therapeutic benefit. So the uptake mechanism of the MIBG molecule is as a norepinephrine analog. So it's very different than the somatostatin receptors that are used for Lutathera."*

The biggest differences are that Azedra is much higher radiation levels, much higher activity, and so the precautions that are needed to deliver the therapy are a little bit more involved. Lutathera is typically done as an outpatient procedure, where the patient receives the therapy during the course of the day, and then goes home at the end of the day. For Azedra, it typically requires an admission into the hospital for about four to five days because of the radiation levels are so high that they need to be isolated. PRRT typically is done with 200 millicuries and Azedra has done with 500 millicuries. And that way also

we can kind of monitor for side effects and any changes during those four or five days. And once the radiation levels are at a safe level, then the patient can go home.

So that's one big difference. The other is that we do a patient specific dose for Azedra, as opposed to Lutathera, which is just a standard dose of 200 millicuries, typically. So what that requires is some pre-therapy dosimetry, where the patient has to come to the hospital they're ultimately going to get the Azedra from, but undergo several scans over the course of one week. And then we take that information from the scans, do some calculations on it, and calculate the specific dose for that patient. Then the patient will come back either the next week or in two weeks, and actually be admitted to the hospital for the treatment.

For patients with pheochromocytoma or paraganglioma, they often can have uptake with MIBG, but also can have expression of somatostatin receptors. And what we typically try to do actually is to do both types of scans and then see which one has the higher uptake, and then choose the therapy based on that."

One big difference between these two options for patients for whom either might be beneficial, is that MIBG is currently FDA approved for Pheos and Para. Lutathera, while it is used "off-label", does not currently have FDA approval for these tumors.

Mittra: *"Azedra is only approved for patients with pheochromocytoma or paraganglioma, and not for other types of neuroendocrine tumors, whereas Lutathera is only approved for patients with gastroenteritis, pancreatic neuroendocrine tumors, and not for patients with pheochromocytoma or paraganglioma."*

Dillon: *"It's not FDA approved for these for pheochromocytoma and paraganglioma, though there is plenty of evidence that it works probably as well as the MIBG therapy and it is recommended by professional organizations for treatment of recurrent pheochromocytoma or paraganglioma. The problem with a therapy that's not FDA approved is insurance coverage for it."*

It's important to remember that neither of these treatments is curative, and neither has been shown to lead to significant reduction in tumor size.

Mittra: *"So the real goal of both of these types of treatment is to just prolong the progression, and survival endpoints. So just to kind of halt the disease from continuing to grow. And typically it's in the range of several months to several years that we see a decrease in the progression."*

This decrease in progression of disease can have dramatic effects in the quality of life of pheo and para patients.

Mittra: *"We have had patients - and I know this is true at many other institutions, as well - who are really quite debilitated by the time they're coming to us for this therapy for a number of different reasons. Either their diseases have simply progressed or, or they didn't have good options to receive certain other treatments, but they come to us and then we've done, you know, one or two cycles of the therapy. And almost immediately they've had a real impact in their lives in terms of improvement in their condition and what they're able to do throughout the day and returning to work.*

We had one patient who was using a wheelchair when they came for their initial therapy and then subsequently had changed to a walker and then changed to a cane and then was just walking normally multiple miles a day, which was just such a remarkable change to see. So it's really satisfying to see that and have such an effect on patient's lives. Even if it's not permanent, that's a major impact."

There are also systemic medical options for treatment.

Sharma: *"There are targeted therapies - tyrosine kinase inhibitors, sunitinib has traditionally been used. There are newer trials with cabozantinib, the multi-tyrosine kinase inhibitors, axitinib, and also everolimus is being tested. Chemotherapy has been used in circumstances also."*

Fishbien: *"And I have to put a plug in for clinical trials, because there are a lot of clinical trials going on right now for patients with unresectable or metastatic Pheo/Para. So it's important to sort of know those options as well because there are new therapies, hopefully on the horizon."*

There's really one treatment decision that someone with a Pheo or Para can make which is more important than any other, and it's this: get your treatment from a care team that specializes in these particular tumors. This is a very rare, very subtle, and very complicated cancer, and working with real experts can make a huge difference.

Dillon: *"These are very complex tumors. It is truly important for patients to find physicians and physician groups that have an expertise and an understanding of the different dimensions of this type of tumor."*

Greenberg: *"When I talk to new patients, a lot of the biggest things that we talk about is how important it is to have a multidisciplinary care team. When we sit down every month at our pheo/para high-risk clinic tumor board, there is a surgeon there and an oncologist there, and an endocrinologist there. Our research team is present. The nurse that helps us run the clinic is there. I'm there as a genetic counselor, and we all contribute to the care of this patient. And it's really important because in pheo/para realm, there are so many different needs, right?"*

If you're going to resect a tumor, you may also need to have an endocrinologist involved for the proper blocking prior to surgery. And that requires both a surgeon and an endocrinologist. So, I think having a multidisciplinary team is really crucial, especially when we think about inherited risks, because these tumors can pop up in different parts of the body. So it's important to have somebody who is a head and neck specialist and somebody who's a urologic specialist and somebody who's a cardiothoracic specialist involved in your care team to make sure that as things arise, you're at a place that has a lot of experience and also is able to talk between the silos of specialties to coordinate care in a really multidisciplinary way.

When something is rare and complex and complicated, you want to make sure that you're somewhere where they know what they're doing, and they've seen it before, right? You want to make sure that when you talk to a provider, you're asking, how often do you do this? Because the more that somebody has experienced in it, the better off that your outcomes are likely going to be."

Perrier: *"This is not the role of an occasionalist. There are just too many nuances and the comprehensive care of the patient is critical. What should I do? What should I take into consideration? What discerning factors for the possibility of following this patient for a lifetime, and for them living in harmony with this disease."*



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