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

In today's episode, we're going to talk about the experiences of adolescents and young adults living with NETs. They are referred to as "AYA" NET patients. It's a group that faces a very specific set of challenges and needs particular kinds of help navigating through the NET journey.

To begin with, what is the definition of a "young adult" when it comes to cancer care? Well, it depends on who you ask and where

you are in the world. Here's Dr. Mark Lewis, Director of Gastrointestinal Oncology for Intermountain Healthcare, which serves patients in Utah, Idaho, and Nevada. He's also a NET patient himself. He was diagnosed with a pancreatic neuroendocrine tumor at the age of 30.

Lewis: *"So, adolescent and young adult patients - actually, the definition varies by country. So, United States has the broadest definition, which is an age range that spans from 15 to 39, which obviously is extremely broad and covers a lot of biology and even psychosocial circumstance.*

And this is a group that's near and dear to me because until quite recently I was considered a young adult, and have now aged just barely out of that category. But when I was first diagnosed with neuroendocrine tumors, I did fit pretty squarely in the middle of what we consider AYA."

Because there is such a broad range of life stages within that under-40 group, we're going to narrow it down for this episode and concentrate on those who are diagnosed at age 30 or younger - people wrestling with the question of what it means to live with cancer as teenagers, or college students, or just beginning their professional lives.

And unfortunately, while NETs remain rare among young people, the rate of diagnosis is increasing. Here's Dr. Manisha Shah, a medical oncologist at the Ohio State University Comprehensive Cancer Center:

Shah: *"There is definitely an increase in the frequency of young adults that I see, male and female, less than 40, less than even 30."*

As with the rise in NET patients in general, some of this has to do with better diagnostic techniques and increased knowledge of the disease among physicians, but that doesn't seem to completely explain the trend.

Shah: *"So, I do think that the part of this increase that we're seeing relates to people seeking more medical care, more active participation in their health, more testing done for one reason or the other, having sensitive tests available. So, that's part of increase, but other part of increase seems a true increase in the frequency of these diseases. I wish I knew the answer why we're seeing the increase so we can work on the prevention strategies, but we don't really know."*

And actually, the number of younger people living with NETs is probably much higher than we realize because AYA patients are especially likely to be misdiagnosed, sometimes for a long time. Here's Dr. Jaydira Del Rivero, a researcher at the National Cancer Institute at the National Institutes of Health in Maryland:

Del Rivero: *"The young adult population, sometimes we don't diagnose it early enough, because it's not like adult where we already have standardized procedures like colonoscopy,*

endoscopy that the patients undergo. But for the young adults or pediatric, it's not that simple."

Shah: *"You know, we wouldn't think 'abdominal pain, oh, is there is something seriously wrong with these patients,' because they are young. The abdominal pain, diarrhea would be labeled as like irritable bowel syndrome or indigestion, some abdominal pain related to maybe the exercise pattern, if they have that. You know, musculoskeletal pain and shortness of breath would be more like anxiety or panic attacks."*

For some of the same reasons that clinicians do not always consider the possibility of cancer when diagnosing health problems in teens and young adults, a cancer diagnosis is a shock for young people.

Shah: *"While none of us want to get sick, when a young person gets sick, it's an extra hard blow because we're supposed to be just living a normal life. We're not ready to have any diseases."*

Here's Kaylen Fletcher, a licensed independent clinical social worker, who runs a support group for young adults with cancer at Beth Israel Deaconess Medical Center in Boston:

Fletcher: *"Fear... denial... how could this be me? I think that's the biggest thing I see is, 'I was just at the gym last week and going on a date. How could this be real?' It's the sense of shock because there's no real reference*

point. Most people in this age group really don't have a lot of experience with serious illness, so it can really kind of feel alienating to be the one, the person who has this serious illness and needing to explain that to people and just feeling different."

Shah: *"They also tend to suffer longer without a diagnosis. So, they have a lot of things to process emotionally to say, 'Gosh, over the last 2 years, I was told that all I have is irritable bowel syndrome, and now I have stage four cancer.' So, this kind of anger or mistrust to the medical system that we cannot blame them for because that's what they have experienced. And so now, to start this new stage where they have to be able to trust the medical system is another layer that comes into the conversation. Fundamentally, they have to know that this is a new step and we want to set in the right direction."*

Let's look at how these emotional and lifestyle challenges present themselves in two distinct groups within the AYA spectrum - starting with that first "A", adolescents.

Shah: *"These are patients who can have disease diagnosed at the advanced stage or a disease diagnosed incidentally at early stage."*

Del Rivero: *"The youngest I have seen here at the clinical center has been 14 years old."*

Lewis: "Teenagers really are, as we want to recognize in society, their own group. I think it's a very liminal phase where you're trying to establish your identity, and it's a really tricky time to be dealing with any serious illness. It can really disrupt sort of how you perceive yourself and how you start planning for your future. And I think it is really knocking some people off their trajectory. You know, I think when you're coming out of high school, you sort of imagine your life being very linear. And if there's anything I've learned, it's that it is very nonlinear. I think that John Lennon said it best when he said, 'Life is what happens when you're making other plans.'

And so, I think it's very common to sort of think of your life in blocks. You do 4 years of college, and then you get a job, and then you start a family. But I think any cancer, but particularly NET syndromes and especially hereditary NET syndromes, can really disrupt that."

One of the many problems teenagers with cancer face is that they can find themselves in a kind of middle space within the medical system, caught somewhere between pediatrics and adult medicine and not completely belonging in either.

Lewis: "Pediatric and adult oncology are different specialties. The line between those groups is pretty thick and impermeable in terms of our training, but biologically it's not. So, age is a continuous variable. You have to choose where you're going to draw your line. And my point

is that many adult oncologists are actually not that comfortable taking care of teenagers, whereas many pediatric oncologists are not that comfortable taking care of young adults. So, the teenagers sometimes kind of get caught in the middle.

Even within the NET community, adolescent patients can have a hard time finding information that applies to them. While NET primaries that are common in older adults, like pancreas and lung, can also occur in adolescents, other primaries that are extremely rare in older people, such as NETs of the adrenal gland, can be among the most common in young people. Even the treatment strategies that doctors apply to NETs are not frequently studied in younger patients.

Del Rivero: *"The way we treat the pediatric population is based on the adult studies. They have some adjustments in the dose based on their height and their weight, and then whether that's effective as we have seen on the adults is something that is a question that to me isn't answered. I think further studies need to be done."*

Some good news about adolescent NETs is that with current care, many patients can live with the disease for a long time.

Lewis: *"I have actually had to conceive my own illness as chronic, something I will never be cured of, but one that still allows me to work, and be productive, and do something I find very fruitful. I'm very, very blessed in that."*

But managing NETs as a long-term condition also presents specific challenges. This includes thinking about downstream side effects that wouldn't present an issue for a geriatric patient. If a treatment might have a serious side effect 20 or 30 years down the line, that's not much of a concern for a patient in their 80s, but it might be a very serious concern for a patient who is 18.

Lewis: *"You have to exist in the moment and cure the person of the disease if you can, but you also have to think about 'how are you affecting this person in the long run?' And it's hard not to sound ageist, but that really is part of the calculus when you're taking care of young people. The average age of a patient in my practice is 68. And so, there's less years of life to protect is one way of looking at it when you're treating an older person as opposed to life years gained treating a younger person."*

This might make a clinician think twice about giving a treatment to a young person that they would have no hesitation recommending to an older one with the same diagnosis.

Lewis: *"I think the one that probably bothers me the most, to be honest with you, is the notion of using PRRT - so peptide receptor radionuclide therapy - in a young person, because we know that there's this small, but again non-zero, risk of causing leukemia. And we don't yet know exactly how that risk plays out over time."*

One argument actually is that it's more likely to happen in older adults who already have less bone marrow resilience than is in kids, but I would be a little bit hesitant using a therapy with that potential devastating long-term toxicity in a child."

And in addition to the long-term and cumulative effects of different treatment options, the cumulative effect of radiation exposure from imaging also has to be considered.

Lewis: *"So, when you get a CT scan, it's a glorified stack of x-rays. That's literally what it is, is x-ray upon x-ray then reconstructed in the computer. So, CT is computed tomography. So, it's putting it all together, really allowing us to reconstruct you in 3D, if we want.*

But a standard CT scan, say of the abdomen and pelvis, is 200 x-rays. And for a long time in oncology, the dominant paradigm had us scanning people every 3 months. So, if you do that over 5 years, that's 4,000 x-rays, which in someone who has already had cancer is potentially enough to stimulate another one. So, one of the things I'm very cognizant of in myself and in patients I'm following over time is radiation burden."

Carrying the weight of all these new challenges, both medical and social, can unfortunately sometimes cause teenagers with NETs to develop an unhelpful or even self-destructive attitude

toward managing their disease. This can take the form of either ignoring potential consequences or wallowing in them.

Shah: *"With young adults, you know, there could be scenarios where they have this sort of a positive attitude, everything is going to be okay, so the sense of taking things lightly. And so, I try to emphasize the seriousness of this disease and the long-term consequences of this disease."*

Lewis: *"I will say I've seen several teenagers who, unfortunately, are prone to nihilism, meaning that they get a diagnosis like this that's really serious and they sort of feel like this is the end for them. Where in fact, we hope that we can give them many decades, not just of life, but a good quality life, where they can be productive and achieve all their goals."*

I often tell patients at the beginning I intend to have a relationship with them therapeutically that will last decades, and I don't want them to fall into the self-fulfilling prophecy of nihilism, where they forego care that can be life preserving because they just assume it's all for naught."

Shah: *"So, I think that is kind of the first thing, to just get on the same wavelength about what these diseases and what its implications can be in their lives and their family and friends' lives. The goal here is to really open up the space where they can be living their life as fully"*

as possible so that we are not imposing limitations. We don't want them to clear up rest of their life to just be a full-time patient and talk about like, "Okay, I have this, I have that, now I can't get married or I can't have travel." Not let cancer cripple you because that would serve no purpose, whether one has 2 weeks to live or 2 years or 20 years to live or longer. And that's what we are here to help them live their life to their fullest potential."

Patient Story #1:

Sophia: *"My name is Ann Sophia Hertado. I am 17 years old. I turned 18 on October 25th, and I was diagnosed in January of 2019, the 31st, with a neuroendocrine pancreatic tumor with metastases in my liver as well. And I live in Sunrise, Florida."*

Mary Anne: *"My name is Mary Anne Hertato. I am Sophia's mother. I am a native of the North Shore of Boston, but we are living here in South Florida, and I am an English teacher at a local private school."*

Sophia: *"Basically, I was diagnosed when I was in my sophomore year of high school. Freshman year, I had symptoms slowly coming on. But sophomore year, mentally, I*

was just checked out 'cause my hormones were so crazy and out of whack that I couldn't even pay attention in class anymore."

Mary Anne: *"You know, knowing what I know now, I realize it started even before that because of the way neuroendocrine tumors grow, but there were little signs. She started her period and then she stopped getting her period. So, I did what I was supposed to do. I took her to the doctor, they did an ultrasound, and they said, 'Oh, everything's okay. You know, let's give it another year.' So, okay. Then there was the weight gain. And I said, 'Well, you know, I'm heavy.' So I was like, "Okay, that might be a little bit normal.*

And then at Christmastime, we were actually up in Massachusetts, and she was just so sick, and she wasn't herself, and she was tired, and she was cranky."

Sophia: *"I am a cheerleader, so I was always very active and I was running at least 2 laps a day around the track field and doing exercises, and I was very social. But there came a point in time where I would get out of breath, I couldn't go upstairs like body weight was just so heavy that I couldn't pick myself up off the ground or get upstairs, and my legs were swollen. My face, I gained weight in my face."*

Mary Anne: *"And my husband went to Nicaragua to see his family at Christmas and then between him going there and us*

going to Boston, coming home, he didn't see her in about 3 weeks. And he saw her at the beginning of January. He's like, 'Mary Anne, there's something wrong. She doesn't look like the same kid she did when I left 3 weeks ago. Her face is huge, and it's round.'

Next day, she went to school. I sent her to the clinic to get her blood pressure taken. It was 152 over 124. And so, it was the high blood pressure that keyed me into there was something drastically wrong. I called the doctor, and I went in that afternoon, and we just happened to get the right doctor at the right time. He took her blood pressure. He's like, 'Oh wow. You need to take her to the ER. She's having a hypertensive crisis.' Okay. And then as I was walking out the door, he goes, 'Tell them I think it's Cushing Syndrome.' I go, 'Cushing? What's that?' He goes, 'Don't worry about it. Very rare. I'm sure she doesn't have it. Just tell them I said that.' And because he said that to me, it made all the difference because when I went to the emergency room and they started looking her over and I said that, they had to look. And that's when they did the blood levels and the cortisol in her blood was so high that they kind of figured it wasn't just a pituitary one. And then they did the scans, and they found out the next day that she had a 16-centimeter tumor on her pancreas.

Listen, when we found out about what was actually involved with this NET tumor and what the prognosis and all of those things were, my husband and I, we prayed, and we just begged God for time. Our decision was maybe if we just have

time with Sophia, maybe they'll come up with a cure, and we begged God for time."

Sophia: *"I get the question a lot of why are you fighting so hard, what are you fighting for, and how do you stay so strong? And I have people calling me their hero. And I'm like, you know what, quitting to me was never an option. Like it never even crossed my mind to give up. And I'm just so thankful for this opportunity and all the support and all the love that I get from people."*

Mary Anne: *"So, we're just going to buckle down and do what we have to do to get more time."*

Let's look now at the situation for NET patients in their 20s - out of high school and just beginning their lives as adults.

Fletcher: *"So, developmentally, this is just a really challenging life stage for folks in their 20s and 30s. There's usually just a lot going on even without a cancer diagnosis. Most people are really discovering their identity independent of their parents and families. They're beginning to make their own money and make financial decisions. They're deciding who they want to spend their time with. And many, many folks are beginning to think about serious romantic partnerships and family planning. So, therefore, if you put a cancer diagnosis into all of this, it's really emotionally confusing and complicated."*

These complications exist on several levels for young adult NET patients – starting with basic questions of logistics and finances. Very few people in their early 20s come to a cancer diagnosis with the financial resources or the experience with the healthcare system that's required to address all the issues they may face.

Lewis: *"One of the huge things to talk about is insurance. Things really change at age 26. And so, again, this is going to be the case for as long as we have the Affordable Care Act. So, under the ACA, you can get coverage under your parents, sort of living under that umbrella, up until 26. And then we've noticed a drastic shift in financial toxicity as people then get into their very late 20s, and it's because largely they're having to acquire their own healthcare coverage.*

And I often feel like it's double jeopardy because a lot of healthcare coverage in this country, as you know, is tied to your job. And if you're too sick to be employed, then it's very, very difficult to secure affordable health insurance. It's almost like these young people sort of get hit both ways. We're seeing that to be an enormous struggle. Some of the treatments we're talking about are hugely expensive. I mean, I often tell patients that if you're just paying market price, PRRT probably costs something on the order of \$160,000 over the 4 treatments. That's money that very few young adults are going to have unless they have a trust fund, and so you can see how

readily treatment for NETs can bankrupt you. And again, it can hit you both ways. It can make you less employable, and it can make your coverage more expensive."

Young adults who are too sick to continue working will often need to explore applying for programs like social security disability benefits in order to support themselves.

Fletcher: *"And that's really challenging for anyone. However, most folks in their 20s have never even thought of applying for disability or taking time away from school or work, and it's really hard to put a pause on those things."*

And perhaps most dauntingly, many people who are seriously sick for the first time are surprised to discover that managing their care is largely their own responsibility. Not only is it up to patients to make decisions about how, and when, and where to be treated, but they are also largely responsible for managing their own medical records and making sure that all the members of their care team have the information they need.

Lewis: *"So, one thing that young patients really have to be very aware of is when their care is being transferred. So again, classically, if you've been a teenager and now you're entering the world of adult oncology, you're probably going to change oncologists. There are some pediatric oncologists that for sake of continuity will continue following that same person, even as they age out of the demographic. But there's also the case where it's almost like passing the baton where an adult oncologist*

like me will sort of inherit a case from a pediatric oncologist. And again, I don't want to put too much of the onus on the patient, but they have to realize in that transfer that one of the things that has to happen is very exact granular information about what treatments they've had before.

Sometimes when I talk to patients, and this happens regardless of age, I'll say, "What treatment have you had before?" They'll say, "Oh, chemo." But that word is meaningless. That's like saying, you know, "I got an antibiotic." It just refers to a class of drugs. It doesn't give me any really meaningful information.

So, actually, one thing that's happened in the AYA space that's been fascinating at least to help with this is this notion of passports, so actually giving the patients either written or increasingly digital sort of ways to track the treatments they've had and sort of make them the ambassadors of their own information."

Shah: *"What I always tell patients is that the patients are my collaborators and I really am very grateful for this collaboration. Many of the ideas, many of the energy that I get to do what I do comes from these one-on-one conversations between patients and myself, and so I'm very grateful for that."*

Added to these logistical challenges are tremendous social challenges as well - learning new ways to deal with the people around you. Unfortunately, interpersonal relationships of all kinds can be stretched, strained, and turned around by a cancer diagnosis, and learning to navigate these changes can take time. For young adults, this often starts with their relationships with their parents.

Fletcher: *"The dynamics that parents and children have like adult children can be really complicated as we know. It's also very common during this time that parents often kind of regress. They see their child, even though it's their adult child, really sick, and they kind of go back to how they acted when their kid was 3 or 4. And I see that a lot where they just kind of want to be all-hands-on-deck, really involved, there all the time."*

The truth is that even as a young adult, you are an adult, and the ultimate decision maker about your care is YOU. This can be overwhelming, but it can also be empowering. Who you allow in your circle and when and for how long is your decision and something you can take control of at a time when it feels like everything is out of your control. If and when you tell your friends, or classmates, or the people you work with about your medical status is completely up to you.

It can be a challenge also in the phase of life when many are solidifying their sense of self to make sure your identity isn't completely consumed by your cancer.

Fletcher: "I think that that is one of the biggest fears of especially younger folks with cancer is, you know, 'I don't want to be that person. I don't want to be the person like, 'Oh, the friend who has cancer' all the time.' And I think for some folks, taking control of that and setting boundaries with friends, you know, 'For 20 minutes of our time today, we can talk about my cancer, we can talk about what's going on with me, but then I want to switch gears because I want to hear about what's going on with you. I want to talk about The Bachelorette from last week. I want to talk about the other things that make me, me,' and realizing that cancer is a piece of your identity and it's always going to be, but it's not all of it by any means. So, making sure to kind of set the tone with friends - with friends and family - can be really helpful, and that takes work. That definitely takes work, and it takes practice."

It's important work, though, because building and maintaining positive relationships with friends and family can be one of the most important things a patient can do to make their life better.

Fletcher: "I have definitely experienced patients who are just like, 'I'm done. I'm just going to kind of hole up and not talk to anyone.' And that's really hard, and it's something that I work a lot with people who just kind of turn inward. And over time, people tend to realize that social support is one of the best tools for getting through a cancer diagnosis and getting through cancer treatments."

With that said, patients tend to figure out on their own who the people are who are going to be most supportive to them, who they're going to want to come to appointments with them, who they're going to want to talk to about their hard days. And there are going to be certain people, especially within a peer group, within a friend group, who might not be able to tolerate that and who might not be the best to go to for all those things. And it might surprise you. Oftentimes, I have patients who are like, 'The people I was expecting to kind of be there and be my go-tos were not, but the people that I did not anticipate to be there all the time were. They were the people who I wanted to come to hard appointments with me and I wanted to talk to about the really tough stuff.'"

An unfortunate truth about cancer treatment is that some of the medications and procedures that might be crucial for saving a patient's life can have serious consequences for that patient's ability to have children.

Shah: *"When one enters into the advanced stage therapeutics, certainly we have several targeted therapies as well as cytotoxic chemotherapies that are definitely high risk for causing infertility."*

Lewis: *"So, some of the drugs that we use in NETs- not many of them, but it's some - are alkylating agents, and it can damage a woman's ovaries and make it almost impossible or*

much more difficult at least for her to have children. So, that actually is a conversation that we often have in AYA oncology, whether we're dealing with NETs or not."

In young adults, this can force really consequential decisions that they might not have been ready to make.

Fletcher: *"Yeah, the fertility piece is really hard because it's another thing that feels like it's being taken completely out of your control. And oftentimes for folks, especially in their 20s, most of my patients aren't really thinking of having kids in the next year or so, but having fertility impacted by treatment forces them to kind of think about that and whether they want kids before they're ready and before they wanted to."*

Del Rivero: *"Am I going to get married? Can I father a child or can I have - If it's a woman, can I carry a pregnancy to term? What are the options? And that's a very common question that I have with a younger patient."*

Fortunately, as hard as these decisions are, there are good strategies available for preserving fertility with a little bit of care and planning.

Shah: *"There are several things one can do in advance. So, the more in advance a patient is informed, they have time to think about it and either if they want to have a pregnancy that they would plan a pregnancy if they're able to because they're not on any medications or that they want*

to have a referral to the specialist for ova preservation or sperm preservation and banking and things of that nature. So, these days, we have a lot of technology that make so much possible."

Unfortunately, these strategies can often be very expensive, so that needs to be planned as well.

Lewis: *"Insurers... And I have to bring this up because there's the practice of medicine, but then there's the business of healthcare, we have to consider both of them. Insurers do not - and you can argue this is shameful - do not universally cover oncofertility, meaning efforts to preserve someone's childbearing potential even when it's endangered by lifesaving cancer treatment."*

All in all, these questions about family planning and fertility can sneak up on a young person who is already overwhelmed by dealing with cancer and can strike them in ways that they weren't expecting.

Fletcher: *"It brings up a lot of emotion. And I think it surprises a lot of people the emotion that it brings up because it can feel like a real loss, kind of this loss of spontaneity around thinking about these things, and it turns into much more of like a medical plan and decision. And it's really hard. It's really hard."*

Questions of family planning can be complicated by the fact that neuroendocrine cancers in younger people are sometimes caused by genetic conditions. And in many cases, these can be passed down from parent to child.

Lewis: *"Neuroendocrine tumors that tend to happen in really young people like adolescents are probably driven by germline mutations, so meaning that they inherited that risk from their parents or it actually happened de novo. So, there are some mutations that your parents don't have at all, and they just happen in you. So, tuberous sclerosis, for instance, is a condition where that actually happens more often than not that the patient is affected, is the first one in their family to be affected."*

Dr. Lewis has a genetic mutation called multiple endocrine neoplasia type 1 or MEN1, which affects a person's thyroid and often causes symptoms related to the way the body processes calcium - some of which can be quite painful and debilitating. In a significant percentage of patients, MEN1 can also cause the development of pancreatic NETs.

Lewis: *"So PNETs in MEN1 are not universally prevalent. And the other word that we use in genetic syndromes is penetrance, meaning that if you have the mutation, how likely are you to have a given condition like, say, PNETs in MEN1? So, hyperparathyroidism is 100% penetrant in MEN1, meaning if you have MEN1 and you live long enough, you will get hyperparathyroidism. The penetrance of PNETs is not*

nearly that high and the estimates vary, but it's probably at least half."

Unfortunately, though, PNETs are the #1 cause of premature death among people with MEN1.

Lewis: *"It used to be that MEN1 patients died of stomach ulcers. They don't anymore, thankfully, because our medicines have gotten so much better. Now, the main threat to life is PNETs, and specifically PNETs that either are hormonally overactive and or PNETs that spreads to the liver. So, the reason I had my Whipple surgery in 2017 is my dominant PNET in the head of my pancreas was growing pretty quickly and pretty large and really posed a high risk of metastasis. And so, I had that removed almost as a preventative maneuver. So far, 3 years later, my liver is clean, but that's why I had the surgery that I did."*

For Dr. Lewis, as for many younger NET patients, diagnosis of both his NET and the underlying genetic condition that caused it came as a surprise.

Patient Story #1:

Lewis: *"When I was 30, I was just starting my cancer training. I had horrible abdominal pain. And showing you the folly of self-diagnosis, I actually thought it was*

appendicitis. But when I went in to get checked out, it was actually my calcium level was very high and what that does is it's almost like the brakes on your gut. So, I had a condition called bilious where my bowel just wasn't moving at all, and that can be quite painful, and that's actually what was setting me off.

So, I'd already gotten through medical school and residency, so I was committed to my profession. I'd already invested a lot of sweat equity and student debt at that point. So, I was pretty convinced that this is what I wanted to do. And I was married. And so, I almost sort of felt like I had sold my wife a bill of goods. Like when she married me, she didn't know sort of what she was getting into. And you can argue the other way too, is that when you already know as a young person that you're dealing particularly with a hereditary syndrome, it does affect your relationships and sometimes your sort of willingness to have a family.

By the time I found my syndrome, I had already had my daughter, and I kind of always wondered in hindsight would our decisions around having a family have changed if we had known before we started having kids that I had this condition because there's a 50/50 chance that I would pass this on to each child. As it happens, that daughter is unaffected, but I also have a son who is affected.

He's 9. His name is Alan, after my father, and he is a ball of energy. And he has zero, at this point, outward signs of

MEN1. And my wife is a pediatrician. And we had a long talk actually, especially once he tested positive, about how to tell him. And it's interesting, I've actually... People love to tell you how to parent. I think everybody has to kind of figure out how to do it on their own, but some people have told us, or told me at least, that we've robbed him of his autonomy because we should have waited till he was 18 and then allowed him to find out this on his own terms. Personally, I would not have wanted to celebrate my 18th birthday with news that I have a genetic condition. And I think the counter argument there is to the extent that we can, we try to normalize this for him in the sense that he understands now when he goes to the doctor, it's slightly more involved than, say, when his sister goes to the doctor.

So, my son really has viewed our condition honestly as something that we share. We have a special handshake that we do, and he calls it our shared tummy troubles, which I find very endearing.

And the other thing is I'm an only child, so if I hadn't had an affected child, this syndrome in my bloodline actually probably would have ended with me. So there is a little bit of guilt, to be honest with you, associated with the notion of transmitting the syndrome on.

I took sort of a huge step towards prolonging my life by temporarily endangering it. So, by doing the Whipple surgery in 2017, it's the largest operation I've had by a

long shot, it was really an investment in my current state of health. So, it was a big, big surgery. I had complications from it, required feeding tubes and a variety of other things for months on end. But ultimately, it's been worth it because now I'm here 3 years later. I feel fine. I do take pancreatic enzyme supplements because I have lost about half of my pancreas and that affects digestion. Other than that, I feel great. I feel able to do the job that I love. I'm very active with my family, and I'm very fortunate. But I also live in a state of, without sounding paranoid, surveillance.

As we come towards the end of this podcast, we'd like to offer three pieces of advice for younger people who are wrestling with a NET diagnosis. First, it may feel like your life is now entirely out of your control, but you might be surprised by how much control and confidence you can take back with a little thought, perspective, and patience.

Fletcher: *"When a cancer diagnosis comes up, it really feels as though all control has been lost, particularly when you're in the beginning of a career, you're in the beginning of a relationship and then all of a sudden this big, huge thing comes up and it's like, 'I have no control over anything here.' So, I think really piecing apart the things, even if they're small, that you can control in a situation... You know, I often have patients make a list of the things in their day they can control and the things in*

their day they can't control. So, getting a certain medication at a certain time maybe you can control. You probably can't. The fact that you have cancer, you can't control. The things you can control are the routine you set for yourself, so the time you wake up in the morning, the individuals that you contact during the day. Really kind of sussing out the specifics of what you do and do not have control over can feel really, really liberating. And also, it's a good reminder that there are things that you have control over."

The second is that you are not alone. However rare your particular condition, there are others who know what you are going through. And because of the internet, there has never been a better time to find them.

Lewis: *"So, I've had several patients recently and heartbreakingly tell me, 'I'm the only person of my age going through this.' And that's another place where actually we can lean into advocacy organizations and support groups that allow people with really rare illnesses to come together and find that, actually, they do have kindred spirits. They might not live in their town. So, when I was diagnosed with my condition, it was a 1 in 30,000 incidence in United States, so I didn't know anybody who had MEN1. So, actually, I found them online. That's how I found my community, and that's where I've stayed in oncology social media for over a decade now. So, I think the other thing to realize in AYAs is they do get the sense that, 'Oh, my gosh, why is this happening to me, and why*

me, and why only me?' And the latter part I really try to mitigate by telling them, 'Listen, you're not alone.' And I think that's hugely empowering."

Fletcher: "Some cancer centers, not all, have young adult support groups. I cannot recommend support groups enough, even for folks that don't think it's for them. I think try it. If you hate it, you never have to go back. Also, most support groups right now are virtual, so you don't even have to leave your space. The Samfund is an excellent resource. They have tons of information regarding grants and other financial tools that can be helpful for folks who are going through cancer treatment. The last one I will mention is Stupid Cancer, which is an organization solely based around young adults with cancer. And they have a conference every year, and this one was actually virtual and recorded, so there are sessions on everything from fertility to body image, to navigating disability, things like that."

The third is do not give up hope. There have never been more successful treatments for NETs than there are right now.

Shah: "I think a lot of hope that I see and I have experienced within our field just of neuroendocrine tumors over last couple of decades that keeps growing is that what has happened in the field about variety of progress in the testing, the diagnostics as well as the treatment aspects of these cancers. So, I would be the last person to tell somebody to say, 'Gosh, okay. Well, these are the only

choices you have.' I tell them that these are the choices that you have now, but I am hopeful that there are more choices coming in coming times."

Lewis: *"So, the silver lining here, and I can't stress this enough, is that the prognosis for many young NET patients is actually very, very good. So, I tell patients my goal was to have a long-term relationship with them. You know, I've moved practices, but I can envision taking care of some of my NET patients for decades."*

Between the time we recorded her interview and when this episode was edited, Sophia Hurtado passed away. Despite her loss, her mother, Mary Anne, wanted Sophia's story to be shared. This episode is dedicated to her memory.

Thanks for listening to NETWise. I'm Elyse Gellerman, and I'm the CEO of the NET Research Foundation. This episode was produced by David Hoffman of CitizenRacecar, assisted by Garrett Tiedemann. It was made possible by the generous support of Ipsen, the Vincent E. Taylor Patient Education Fund, Advanced Accelerator Applications, a Novartis Company, and Lexicon Pharmaceuticals. Special thanks to everyone we interviewed for this episode. We are grateful for your expertise. This is a production of the Neuroendocrine Tumor Research Foundation where we're committed to improving the lives of patients, families, and caregivers affected by neuroendocrine cancer by funding

research to discover cures and more effective treatments and providing information and educational resources. Please visit us at NETRF.org

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