Transcription: “Primary immunodeficiency in 2026: New practice parameters, genetic testing, and what's changed for patients”

If you or someone you love is living with the primary immunodeficiency, you know that the journey can feel isolating. But this June, you can see for yourself that you're not alone. Join us for the world's largest gathering of the primary immunodeficiency community at the immune deficiency foundation's twenty twenty six PI conference in San Antonio, June twenty fifth through the twenty seventh. Whether you're newly diagnosed, a long time advocate or a family member, your story matters. From life changing educational sessions to meaningful new friendships, this is where our stories come together to form a beautiful mosaic.
Head over to primary immune dot org slash conference to register now, and we'll see you in San Antonio. On this episode of the Immune Deficiency Foundation podcast, doctor Paula Henao and Katherine Lontok Ph.D., the foundation's medical director and director of science and policy communications respectively, discuss the highly anticipated practice parameters for primary immunodeficiency or inborn errors of immunity. The new parameters intended to guide clinicians in diagnosis and treatment decisions are more comprehensive and holistic than ever before and include topics such as genetic testing, newborn screening, mental health management, and much more. Let's get started.

Paula Henao: So I I wanted it today for us to just sit down and chat about these new practice parameters that just came out. And for for everybody in the audience, I'm Paula now. I'm an immunologist and associate professor at Penn State Medical Center. And I am the medical director of the immune deficiency foundation, a foundation that's a patient advocacy organization that helps us give the voice of the patients that have immunodeficiencies. And so and, Katherine, it's so nice to have you.

Katherine Lontok: Thank you. I'm Katherine Lontok. I am the director of science and policy communications also at the new deficiency foundation. And we've been waiting for these parameters, so it's very exciting that they are finally out, and they are they are meaty. There's a lot that

Paula Henao: They are meaty. That oh my gosh. Yes. So it is over fifty pages and I believe eight hundred thirty seven references.

Katherine Lontok: Very extensive.

Paula Henao: Very extensive, very well researched. And one thing that I did want to pause and give a huge kudos to the writers, editors of the parameters, Jordan Orange, and his team really took this one out of the park. I'm not always are the practice parameters, so

well thought out and just give the information that is most necessary in the field. And this is a heavy one because it includes diagnosis and treatment of IEI. So I don't know, Catherine, what are your thoughts about what are the parameters?
Why why do they matter?

Katherine Lontok: Yeah. So practice parameters and the reason these are these are so meaty. Is they're tools, right, for clinicians, but also the field as a whole, so that everyone's kind of on the same page about how you diagnose and treat and manage, you know, particular medical conditions. And so this set of parameters is all about primary immuno deficiencies, also known as inborn errors immunity. And the great part about these parameters is they they not only give recommendations, but they actually grade the relative strength of the evidence behind each recommendation and the certainty of the recommendation itself.
And the certainty is a little different. That's like what everyone you know, does this a recommendation apply to all patients or some patients? Right? So, yeah, there there's a lot in there because it really is the state of the field.

Paula Henao: Yeah, absolutely. And these were a joint effort from professional organizations, the AAAAI, the College of Allergy, and the Clinical Immunology Society. Altogether created these parameters in a team effort. So when we create these parameters for those of who are not familiar with this, basically, the leads in the field meet up, they discuss, and therefore, are able to figure out and confer what is the main recommendation. Some some areas are very easy to recommend, but others are heavily debated.
So what comes into the parameter is a deep effort, deep debate, and it comes from the leading minds in the field.

Katherine Lontok: Right. And these these particular parameters are, you know, they were last updated in twenty fifteen. And you know, have just now been released in twenty twenty six this update. So there is an enormous amount of research and discovery that's happened in the intervening years that these parameters have to try to address.

Paula Henao: Yeah. One thing like, the parameters really have made a lot of big changes from two thousand fifteen. And and it's understandable that so much has happened since then. But what are what are just broad strokes? What are the areas that you think have been the biggest, most impactful changes in the parameters from twenty fifteen to twenty twenty five parameters even though they were published in twenty twenty six.

Katherine Lontok: Yeah. Yeah. I think one of the big things is the real focus on mechanism and pathology. So really trying to connect what patients experience their symptoms and signs to what what is the variant at the DNA level? And how is that affecting, you know,

things all the way up to the whole person?
And so that emphasis means for diagnosis, right? There's a really big focus on genetic diagnosis and using the genetic diagnosis then to actually guide treatment and management. Yeah. And what else do you see, Paula? I'm sorry, you picked out something else.

Paula Henao: I think that there's that is that is a big one. Right? The the focus on genetics, which wasn't there in two thousand fifteen. In two thousand fifteen, it was an adjunct Right. And and in chosen chosen scenarios, And seeing the evolution, I mean, there are conditions that didn't exist in two thousand fifteen in in the parameters.
So one is a p d s. So that is activated PI3K delta syndrome. That was not included in the twenty fifteen parameter, and it was in part because they had just been discovered. It was first described in twenty thirteen, then a couple of their papers in twenty fourteen, and they identified this this particular gene. Well, on behold, It's only been ten years later, which really isn't that much time, and we've gone from identification of that gene all the way to therapy.
So huge big strokes in terms of now we're able to look at a gene that didn't even exist in two thousand fifteen and now have not only a great identification of the gene using it regularly in genetic screening, but not but also targeted therapies, so big things. And also I love that the parameters are thinking about holistic care. They're recommending multidisciplinary care. They discuss the importance of multiple physicians being involved and understanding that it's not just an immunology. Care.
Many of these patients have many issues that need an interdisciplinary team and that they need regular monitoring and expert care. So that's that's important as well. And the view overall that newborn screening, which wasn't universally there in twenty fifteen, is now available in all fifty states, and it's paying off. It has truly saved lives.

Katherine Lontok: Right. Yeah. There's now a whole section in the parameters on, okay, you've done you have a positive newborn screening result, but it's not SCID, now what? Right? And that wasn't really a consideration in twenty fifteen.
Yeah. Absolutely. And

Paula Henao: for those listeners, just in terms of the sheer number of genetic conditions, More than two hundred genetically defined immunodeficiencies back then were defined in the practice parameters in twenty fifteen. And now we're recognizing more than five fifty, and it keeps moving forward. So we're moving that genetic understanding significantly forward in the field. Important to note, Katherine, the recognition of the name inborn errors of immunity. And I know it can sometimes feel as a patient when there is some fears about being labeled something that doesn't feel great.

Right? That the important areas of immunity clips down to some people as, oh, there's an error in me, and I'm perfect, because each human I recognize is innately perfect. Do you have a thought about why that shift in name from primary immunodeficiency to inborn errors of immunity? And how can that have helped advance the science forward? Or not?

Katherine Lontok: Yeah. So the term inborn errors of immunity is actually derived a little bit, at least, from another field, so inborn errors of metabolism. And I think that the goal in switching from primary immunodeficiency to IEI is actually to be more inclusive of the types of signs and symptoms that people have. So there are, you know, a number of conditions we know now, right, where actually infection is not the main symptom. Main symptom is actually, you know, autoimmunity or inflammation.
And the term immunodeficiency doesn't really describe those very well. Right? Because autoimmunity is like your immune system is gone gone off the rails, not that it's Yeah. You know, weakened. Right?
So I think that the the term inborn error of immunity is is an attempt to actually describe the entire spectrum of conditions.

Paula Henao: That's so important too because so many patients and members of our community are not just having issues with infections, but they're having issues that are much more than that. And the infection, sometimes when they we start treating the infections with, let's say, immunoglobulin replacement, the infections become the easier part, and it's the autoimmunity predisposition to malignancies, all of the other areas that become the harder part to deal with.

Katherine Lontok: Yeah. Exactly. The so one thing that I noticed in these parameters, and we've kind of touched on it a little bit, but genetic testing is now really front and center. So it is, you know, these parameters recommend that genetic testing be part of the initial workup. So if a clinician has a patient that they suspect, has an IEI that they should be doing genetic testing.
Even even if at the end of the day, the genetic testing doesn't, you know, pull anything up. And I think that is definitely that's definitely a new emphasis. Absolutely, absolutely. I think there's a lot there because one of the fears about genetic testing is cost.

Paula Henao: And and I think that's still a valid concern. Cost can be insurance can still be an issue with with genetic testing, though, I think that is rapidly changing given the things like this the parameters saying genetic testing is front and center is only helpful for getting insurance companies to pay for necessary genetic testing. Right. And I, for instance, as a clinician, if that is denied, I can show them print them the fifty pages of the parameter and fax them over or mail it to them and tell them, hey, read through this Genetics is super important in the diagnosis of IEI, so that really can be helpful. And and the constant general

of genetic testing was very high in in two thousand fifteen compared to now.
It's

Katherine Lontok: Right.

Paula Henao: It's in lab levels has the actual doing the genetic testing has dramatically decreased in cost making genetic testing a lot more re simple. And more and more in training, I think when when I was training or for as an as an immunologist, genetic interpretation wasn't that emphasized in my training. But now our fellows are learning as part of the their training to really get genetics and and evaluate them and interpret them and think about when do we do genetic testing for a single mutation in the case of I know I strongly suspect it's this particular issue. I'm gonna test for this particular issue. When do you do panels of immunodeficiency?
When do I need to go deeper into whole genome sequencing? So the knowledge of what to do and when is really emphasized in fellowship education in ways that it wasn't in previous generations, which I think is is really fascinating. And and it really shows that in the past when I was in med school, when we thought about genetic testing, it was sent to a geneticist. That was the default. We have to send everything to genetics.
And turns out there's no way that that we have enough geneticist in the country to be able to send people to genetics anymore. We have to each field that is doing genetics has to be able to interpret their own genetics. And the geneticists are an additional resource in certain scenarios, but they are they can't handle the volume of every patient getting genetic testing, but because they would literally be seen half of the population. Maybe I'm exaggerating, but it could get there.

Katherine Lontok: Yeah. Yeah. I actually heard an interesting factoid and, you know, take it with a grain of salt because I haven't followed it up or anything, but the amount is actually now cheaper to just redo genetic testing instead of storing the data and reanalyzing it. Really is yeah. It's really interesting.

Paula Henao: So so, like, they don't they don't wanna like, the genetic places aren't aren't storing. They're just saying, hey, I'm gonna dump it. And then if you need it in the future, you get swabbed again.

Katherine Lontok: I don't know if that's actually happening with the cost for because the data is enormous. Right? It's a huge amount of data. Right?

Paula Henao: Yeah.

Katherine Lontok: And so the cost to store that data is actually more at some point. Right? At some point in time

Paula Henao: Yeah.

Katherine Lontok: Than it is to just redo the analysis, right, from scratch. Yeah. So it's it's I found that to be pretty interesting.

Paula Henao: That makes a lot of sense. And, you know, you and I were recently at CIS, and I cannot remember which talk mentioned that it's reasonable that with with how the advances have gone, that it's reasonable eighteen months later to go back and redo genetics in some of these patients. So I felt terrible. I had not been going back. And I have all my genetics studies, all my patients I'm now taking the task of reviewing every genetic test that I've ever done, which is a huge task.
I have a fellow helping me going through and seeing which ones need to be repeated because we didn't really grab a jeans. And going going forth and having it have asking the genetic lab to to retest. So so I'm I'm doing that as a little perk that I took with me from CIS. That was that was very helpful. I do have a little every time I read a parameter, I struggle with what I did in the past that was wrong because my how many people did I say, hey, your immune system is totally fine in twenty seventeen, whereas now I would have done something completely different.
And that that can be really ten like, cause a lot of tension, things that you know, you do five years from a go or very different from what what they will be five years from now. And and it does tell you in terms of a patient if you have something and it doesn't go away and it gets worse. I don't think it's wrong to go back to your doctor several years later and hey hey, can we reevaluate this because even if you go to the same doctor, they might be having a whole new set of tools, a whole new set of ideas of what they can do for you. Right. Right.

Katherine Lontok: Right. Right. Yeah. And and I think that you know, patients are getting happier at this, right? And even in my own personal life, you know, I've I've had conversations with family members where I'm like, no, no, no, you need to go in and look at the lab results for your doctor looks at the lab results.
So you know, what's high? What's low? What do you need to ask about? Right? So I think being informed and being on top of it and under standing that doctors are people.
Right? So, and this idea of medical knowledge changes over time. Right? So, yeah, like you said, it if you are continuing to have symptoms that are unaddressed, right? You've got to keep going, right, because there there is an answer somewhere.

Paula Henao: And make sure that you're going to professional. I think I worry a little bit about a lot of my patients lately going into different AI platforms for medical information. And my my worry there is that I have to remember that the AI one, it's very agreeable. So so if you want it to give you an answer, you can you can get it to give you the answer that you want potentially. So so that worries me a little bit.

And the other the other part of it is the possibility of having misinformation or just kind of what what sometimes happens is I'm going to give you a broad differential of everything that could be the case then. Yeah. And you if you don't have the background, you could get very lost in that. So, well, it's really important to be your own advocate take a lot of the information with a grain of salt, go to really good, solid platforms, go to the immune deficiency foundation website, that is a website you can trust and others. So that you could get the right information rather than looking in in areas that could be a little bit tampered with social media, things like that.
They they can they can spread misinformation and cost undue anxiety and take you down a rabbit hole, trust your doctors and the reliable resources that have done their work.

Katherine Lontok: Right. Right. So Getting back to the genetic testing where we went down a rabbit hole of our own a little bit. So I think I think it's really interesting again, that it's now so front and center. And I think part of that is a better understanding of the ways that genetic information can impact patients, right?
So it's not just, I mean, APDS is a great case study of. Here are people who used to get lumped in with CBID. And now, you know, because their condition was defined better, defined genetically, there's an FDA approved treatment just for them. Right? It specifically targets the issue with their immune system.
And that's great. But even when there isn't an FDA approved treatment for a specific condition, genetic testing can actually give you a lot of information. Right? They Absolutely. It can it can alert your care team about things that they need to be thinking about.
Right? Like, oh, you have a variant in a gene and patients with that variant tend to develop lymphoma over time. Right? You need to be screened for that and I need to ask you questions at every encounter that could, you know, uncover symptoms. Right?

Paula Henao: Absolutely.

Katherine Lontok: And also, you know, if you understand the genetic basis of something, then that gives you information, not just about the patient in front of you, but potentially their family. Right? And perhaps uncovering other individuals in their family that may be affected and don't really know it.

Paula Henao: I couldn't agree with you more, Katherine. And when I counseling and I've heard many physicians throughout the years and of course this is an evolving science, but say, well, it won't change management. So what's the point of genetic testing? And when I counsel patients, I I do let them know, especially patients with CVID where a genetic diagnosis is is is not the majority, but the minority. And having a monogenic cause, meaning one gene that explains everything is is not always the norm.
Sometimes there's like multiple genes acting up and it's a little bit a more complicated

story. And so I do say, hey, like, the genetic the the genetic testing may not give us an answer.

Katherine Lontok: Right. Right.

Paula Henao: It may not give us an answer now. It may give us a gene, but a gene that we can't act on, like you mentioned, we can't treat. It may give us some information, but it's only partial. But what really also is important to know is it may not give us those that information now. But if we have a gene that we really can't do much about today, at least we know the gene and we're empowered And ten years from now, the scope could be so different, but now you know the gene and you could be like, wow, the the drug or the the research for this gene is coming out as being done right now, I can enroll myself in this research study that's doing this new something for this particular gene.
And that is power in its own way.

Katherine Lontok: Right. Exactly. It's yeah. It's like sort of a positive feedback loop, whereas the more people who have a genetic diagnosis, the more you can approach treatment as well we know this pathway is affected, let's try this drug. Instead of kind of a haphazard.
We don't really know what's going on. Let's just try a bunch of things and see if something works. Yeah. Yeah.

Paula Henao: And and speaking

Katherine Lontok: What's a big sort of sea change?

Paula Henao: Yeah. Absolutely. Speaking to that, the parameters say about five to ten percent, although that could be changing Yeah. Of PI patients have multiple genetic conditions that could be affecting their immune system. Right?
That's not just the one monogenic cause, like, a lot of skid is one single gene being affected. And what are your thoughts of that? And is this – was that new information for you?

Katherine Lontok: I mean, as a non clinician? Yes. That was actually new information for me. What I what I thought was really interesting is not just that number itself, but the fact that there's evidence that people with PI are more likely to have multiple genetic conditions than people with other types of genetic disorders. Right?
So it was something like two or three times as many people with PI have multiple conditions as Yeah. Other, you know, people with other genetic conditions. So and again, it's, you know, it's one of those observations that's like, that's a big question mark. Like, why is that? Yeah.

Paula Henao: Yeah. I think the IEI world has a lot of different factors that are involved. Right? There's the autoimmunity piece, the immunodeficiency piece. There might be just multiple genes that one one hit, one gene being defective may not be enough for the whole for for a for a specific phenotype or specific characteristic, but having multiple genes affected will cause that that error to manifest itself.
So it is it is fascinating that we have more information about these multi gene issues and will create a big question mark and big research opportunities in the future in terms of how to manage those because we've gotten very sophisticated about thinking about monogenic one single gene. But we're not quite there in terms of how do we address multiple genes affecting one one diagnosis.

Katherine Lontok: Yeah. Yeah.

Paula Henao: So one controversial topic that I think you and I have discussed in the past and that has been popping up a lot in conferences has been the diagnosis of specific antibody deficiency. I know the practice parameters do divide into two different diagnoses specifically Mhmm. Specific antibody deficiency related to polysaccharide and specific antibody deficiency related to absence of protein antibodies. And so would you be able to tell us a little bit more about the evolution of that? What's happened with this condition? And and why is it reflected a little different in these parameters?

Katherine Lontok: Yeah. I mean, so the the specific antibody deficiency to polysaccharide antigen. That's the phrase that's used in the parameters is what we traditionally think of as specific antibody deficiency. So these are people who they have normal IG levels, and they and it looks like, you know, that they they should have good protection against many different terms. But they don't make antibodies to polysaccharides, which make up this coating on specific species of bacteria.
So that's what we traditionally think of as specific antibody deficiency. And so this new category specific antibody deficiency to protein antigen is interesting because the idea there is that, again, these folks have normal IG levels, but they have trouble making antibodies to protein antigens. So if you give them a protein based vaccine that's highly immunogenic, which means that basically, everyone responds to it. Mhmm. They don't respond.
Mhmm. So that's I think it's a it's an attempt to categorize a specific group of people that have been uncategorizable, I guess Mhmm. Up until now. So it'll be interesting to see how that evolves.

Paula Henao: I I do think it's interesting because I do see that these are patients that are potentially somewhat different in their in their presentation. We would expect a person that has poor response to, let's say, the tetanus vaccine to typically present us significantly

again, not all patients read the book, but more sicker than than a person with defective polysaccharide vaccine response. So it is interesting in terms of how they made that distinction. And maybe to think about, not necessarily, but it helps us think about maybe treatment is a little bit different. Potentially we could consider IG therapy a little bit sooner in the patients that have more of the protein based vaccination issue compared to the polysaccharide only vaccination issue, although every treatment is going to be patient dependent.
The other thing that they did, which I thought was very helpful, was that they change the cutoff of the percentage from

Katherine Lontok: of

Paula Henao: why the polysaccharide vaccine response is considered protective. So it used to be you you basically, as an immunologist, you'd love you get all the different polysaccharide vaccine titers. Typically, it's twenty three and you count them and the number that are protected used to be greater than seventy percent used to be considered protective. And now it's greater than fifty percent which means a few less patients will be will fall into that category of specific antibody deficiency. They also made some adjustments that are related to the pneumonia vaccine, the pneumovax twenty three becoming less available in the U.
S. Because that's the polysaccharide vaccine and the pneumococcal vaccine in Prevnar twenty and CAP vaccine which is a twenty one pneumococcal anodges are both contingent vaccines, which means that they function essentially as a protein based vaccine. So what they did is they changed, they they said, hey, we couldn't use those vaccinations, the CAP vaccine, and the Prevnar twenty, and those are the conjugate vaccines. But the cutoffs for what is going to be considered a protective titer is going to be different. Mhmm.

Katherine Lontok: Mhmm.

Paula Henao: So that's those are some changes in the parameters, which I think are are relevant for for common practice.

Katherine Lontok: Yeah. Absolutely. So we touched on this at the beginning about newborn screening, right, for SCID that has expanded dramatically. And now, there's a whole section in these parameters on what to do if you have a newborn screening that comes back positive, but it's not skid. Right?
So what does that tell you about, you know, where newborn screening is headed? Where it's come from, I guess, and where it's headed?

Paula Henao: Yeah. So, new of our screening, huge asset to our field. We really have made remarkable strides. It's available in all fifty states as we talked about. And it really is

speaking to that a lot of these patients are screen positive but don't have either skid or congenital athymia.
And what do we do with with those patients and I think a lot of the field was unsure of what necessarily we needed to do, which is why we have to have it in writing and make it clear for people that may not see a huge volume regularly. And so they've really made it very clear. What are the steps that need to be taken? What is the monitoring that these patients need? And what what are the diagnoses to consider when it's not skid or Mhmm.
Congenitally athymia. So that can be very helpful.

Katherine Lontok: Yeah. And I think this this recognition speaks to, you know, when when skid newborn screening was rolled out, I'm not sure that we totally understood what it was going to catch in addition. Too scared. Right? Now we have that information.
Right?

Paula Henao: Yeah. Absolutely.

Katherine Lontok: And I know just in the newborn screening world in general because it is a public health initiative, there is a lot of concern about if we expand it to cover even more diagnosis you know, how are we gonna serve everyone that that screens positive? Right? And, I mean, certainly, that's a valid concern. But I think that SCID newborn screening shows that you you have to you have to start screening to know what you're gonna find and how you need to adjust you know, the whole medical system basically to accommodate these patients. Because it's not like if you don't scream, the patients don't exist.
They still exist. Right? You're just not you're not catching them early enough. Necessarily to do something. So yeah.
I think you have newborn screening and and just the fact that, again, there's a whole section in the parameters now about what to do with the non skid positive screens really speaks to that.

Paula Henao: Yeah, absolutely. And and the future of newborn screening is very expansive. We we we don't know where newborn screening is headed. There's a lot of opportunity their the newborn screening may include something called KRECs, which would diagnose not what KRECs are doing, which looks at t cells, but KRECs look at B cells. And eventually, maybe genetic screening could be a broader incorporation of newborn screening that could help find treatable diseases at a broader range and bring these patients into the healthcare center early for for therapy.

Katherine Lontok: Yep. And then, again, like I said, you know, it's not that they don't exist. We just, right now, we're not catching them. We're not measuring who has what and and, you know, how the healthcare system needs to grow to treat them. So

Paula Henao: Absolutely. Absolutely. This has been such an interesting conversation, Katherine. I'm wondering one thing that is really relevant to our audience because the vast majority of our community although although they have all sorts of issues with primary immunodeficiency and we represent a very wide community and we're very proud of it. The vast majority of our community are people that have to use immunoglobulin replacement of different sorts.
And I'm I'm just curious how did these parameters change if if at all? The administration and recommendations of IG therapy?

Katherine Lontok: Yeah. I found it really interesting that the parameters this time around actually put a number on the serum IgG level

Paula Henao: Mhmm.

Katherine Lontok: That patients that are on IgG therapy should be maintaining. So the actual level that they recommend is at least eight hundred milligrams per deciliter. And the previous parameters did not specify a number at all. And I think putting a number on it can be good, but it could also it could also be a little bit detrimental, which is probably why they haven't put a number on it previously. Right?

Paula Henao: Mhmm.

Katherine Lontok: So you mentioned before having insurance cover genetic testing and you, you know, you can now point to these parameters and say, look, it's a first line test. You need to cover it. Well, the same thing goes with this, right? Somebody who is struggling to get their insurance to cover a higher dose of Ig, they're having a bunch of breakthrough infections. If they can show my serum IgG is dipping below this, I need a higher dose or I need to have a more frequent infusion that's pretty good justification.
Right? For them for going to the insurance company and saying you need to approve this. I think that the the danger comes in because we know that there are people who even act eight hundred still have breakthrough infections and would do better with an even higher serum IgG level. Right? And so the sort of double edged sword there is the insurance can say, well, yeah, but you're at eight hundred, you should be fine.
And ignore the fact that there is a spectrum. Right? Yeah. So I don't know if you wanted to comment on that as well in your experience.

Paula Henao: And I can I can imagine and maybe maybe completely wrong, but that the this was a source of debate Yeah? When when this was this number was decided. So we know that the incidence of pneumonia was fivefold higher with trough levels of five hundred compared to a thousand. There is good reason that higher is better. In common variable immunodeficiency, the data doesn't show that going above a thousand is necessarily in a

population, necessarily gives us better pneumonia protection.
But it there's there's still data showing maybe nine hundred is better than eight hundred. And so so this number eight hundred, I think, was chosen because it was between the some of the data and the meta analysis looked at. Somewhere between eight hundred and a thousand showed less pneumonia. And so that was that was the magic number chosen. But for eight hundred, I think it's nice for me to have a number that I'm aiming for to start with.
Mhmm. Certainly, these IG levels are irrelevant for people that are on immunoglobulin replacement therapy for something like specific antibody deficiency where they already have normal levels and we're just gonna go Yeah. Much higher. So so that would not be relevant there, but I think it's nice to have a number and helps us have this higher goal because eight hundred is a is a reasonable, good goal that is evidence based?

Katherine Lontok: Yes. Exactly. It's just, you know, the insurance companies like to have hard and fast rules for everyone. And it can be frustrating, right? When you are that patient that's on the tail end of the bell curve, right, that to have insurance push back and say, well, but you meet the number.
And it's like, what's Okay. But I'm an individual. Right? I'm not I'm not the average. I'm an individual.
Yeah. And so I do think the fact that it's that it is a relatively high number means that having this number is actually gonna be helpful for most people rather than detrimental. And I just I hope that the insurance companies pay attention to that phrase. At least at least. So at least is very important.

Paula Henao: Yeah. Absolutely. And and going back to the level the the what you were mentioning about insurance, the practice parameters does give a recommendation or for route of administration. And we've been dealing with some insurance issues about an an insurer provider selecting and dictating that only one IG therapy is available, and the practice parameters say something very different. So so I don't know if you wanna tell us a little bit

Katherine Lontok: about that. Yeah. So, you know, there's now written into the parameters, a recommendation that the route of IG administration, so this it doesn't specify the product, but the route. So intravenous versus subcutaneous, be based on patient tolerance and preference. And I, you know, I don't think this is new, this is not a new development.
I think since there's been the option of IVIG versus SCIG, you know, the consensus has been, it really needs to be an individual decision taken into account many different factors. Right? But to have it in the parameter stated like this is really, again, I think, helpful

because, you know, it's it's pushing back on this idea that there is one thing that works for everybody. We know that's not the case.

Paula Henao: And it really does give physicians who are experienced, they know what they're doing for their patients and patients the choice of choosing the product that's going to best treat their condition. I thought just as an added a thought. One thing that I wanted to share was that the practice for Amino's also included that The IgA level was irrelevant in terms of receiving product. And I thought that was really helpful as one of the common question that I get asked by patients, but also by when I'm answering the ask IDF, can I get IG product? Can I get blood transfusions if my IgA is low?
And the practice parameter says, Yes. We don't see in a population basis concerned for people with low IgA receiving Ig product. So I thought that was very helpful.

Katherine Lontok: Yeah, definitely.

Paula Henao: So they also go into a little bit about mental health, fatigue. What was your takeaway in terms of quality of life recommendations from the parameters?

Katherine Lontok: Yeah. So again, I think that this plays on the idea that these parameters are more holistic than they have been in the past. And it doesn't it's not just holistic in terms of having a multidisciplinary team for physical symptoms, but the fact that mental health, quality of life, perceived health, for example, are all things that matter. Right? In terms of a person's overall health.
So, you know, there's a recommendation now that people be assessed for their mental health and their fatigue at every visit. Be interesting to see if that actually happens, but I think, you know, it's definitely a nudge in the right direction. Right. And this, you know, better understanding that all of these things come into play. Right?
A person is a whole person. They're not just their immune system. They're not just Mhmm. You know, one one condition versus another. So it was very nice to see that.

Paula Henao: Yeah. Yeah. But it seems like we're out of time. I wanted first, this is so exciting to be able to talk to you about the practice parameters and about some of what's going on in our community. I wanted to think, again, the writers, the editors, the tables are beautiful,

Katherine Lontok: but Oh, yes. A row. Yes.

Paula Henao: Some of the figures are just chefs kiss kudos to them. It was it's a beautiful piece of work. The work that went into it, I It's it's ginormous, and they're really setting the bar high in terms of what our feel needs to work on and we're we're in helping us direct the future of our field. Wish that they could do this more often because I love them. So thank you thank you to all of the editors and contributors to the practice parameters.

Katherine Lontok: Yeah. I wanted to just say as a as a, you know, a communicator, right, a health and and medicine communicator, those visuals and tables at the front are just so well done. Right? You could print those out. I mean, not that I recommend this.
You should read the whole fifty pages. Right? There's a lot of really good info there. But if you print those out and hang them up in your office as a clinician, right, you're gonna have a pretty good head start on actually following those parameters. Right?
So that's just fantastic. In terms of

Paula Henao: I have been printed and not in my in my in my clinic. So I recommend it. They are beautiful, colorful, very well done.

Katherine Lontok: And very clear. Right? Very very clear.

Paula Henao: Yeah. So thank you to everybody and thank you to everybody that's out there listening. If you have questions, we always have our AskIDF and let us know if you have any other thoughts for future podcasts.