0:06
Hi everyone.

0:07
Thank you for joining today's program and ... featuring our guest speaker, doctor Sammy Calla. We're excited to have you here with us.

0:16
A special thanks to our generous sponsor for today's program, CSL Barry.

0:21
During this session, we're going to discuss different ways a patient should manage the condition.

0:26
From one a patient seeks diagnosis to possible treatment options.

0:30
The program will also highlight how chronic inflammatory ... Poly neuropathy, also known as ...

0:36
varies from other forms of neuropathy.

0:40
My name is Lindsey Kober, for those of you who don't know me, I'm the Executive Director of the Foundation for peripheral neuropathy.

0:47
And then with this organization for over seven years now. And it's exciting that we finally been able to put together a program on ..., our first, ever one on this topic.

0:56
So thank you again for joining us.

0:59
Before we get started, just a few housekeeping: housekeeping items are being shown on your screen. Yes, we are recording this program.

1:07
That recording will be permanently housed on our website for future viewing needs.

1:12
Those that are not participating in today, today's live session will be able to go on our website, and those that are, will get a recording of it e-mailed to them within the next day or two.

1:23
We will be holding a question and answer session at the end of the presentation.

1:28
All questions can be submitted through the questions box in your dashboard.

1:32
And for those of you who are watching the recording later on, I encourage you to ask any questions to the foundation for peripheral neuropathy via e-mail or phone. And we'd be happy to still answer those.

1:43

And now, I'm pleased to introduce today's experts, doctor Sammi kala.

1:47
Doctor Keller has more than 20 years of experience in diagnosing and treating patients with a variety of neurological diseases and is an intending attending neurologist at Penn Presbyterian Medical Center.

2:01
He co-founded the pen Amalea Dosa Center, one of the largest multi-disciplinary programs in the United States treating patients with hereditary and acquired amyloidosis.

2:11
Is other interests includes treating patients with acquired inflammatory neuropathy is such as the IDP and myopathy? Yes.

2:19
And now it is my it's my distinct pleasure to officially introduce doctor Keller and let him take it over from here. So doctor Carla, thank you again for joining us today.

2:31
Thank you, Lindsay.

2:32
This is a wonderful invitation and I appreciate you very much your team and the foundation for peripheral neuropathy and for CSL bearing for putting, for hosting this and or supporting this, I should say.

2:48
Millimeter.

2:49
Let me start by talking about what a peripheral neuropathy is and then we'll, we'll be spending the rest of the time talking about chronic, inflammatory demyelinating poly neuropathy.

3:01
It's a mouthful, and hopefully I'll be able to parse it out for you a little bit, tell you how we make a diagnosis, and tell you the treatment options that we have available to us today.

3:16
So first of all, peripheral neuropathy means disease of up of the peripheral nerves and peripheral nerves are the wires of the body.

3:25
If you will, they are the organs that carry information from our skin, from our joints, from our muscles, up to our brains, so that we can, our brain can interpret what it is in the world around it.

3:45
And then the brain sends back information to the muscles too, make them work. So when you want to move your arm, you want to move your leg.

3:59
There's a lot of signals that are going back and forth across these peripheral nerves.

4:05

And these nerves, of course, are, come out of the spinal cord in our neck and our thoracic and lumbar spine. And then they go to our limbs to our face to are all over the place. Very complex system.

4:23
And most of the time it works beautifully when there is a problem.

4:28
We get the symptoms of peripheral neuropathy, so and they tend to be and fairly common and consistent no matter what the cause of the neuropathy is.

4:43
So you when you damage your peripheral nerve, you get numbness, you get tingling, you get loss of feeling.

4:53
You get pain. That's electrical kind of pain.

4:58
And then you can get muscle weakness where the muscles don't contract very well or at all, depending on the degree of injury to the nerve.

5:09
And, uh, when this happens, depending on the degree of nerve injury, depending on the cause of nerve injury, the nerve can recover to some extent, or it may not. And again, that all depends on how the process of injury has happened.

5:32
So that's basically peripheral neuropathy symptoms.

5:39
Overall in a nutshell, peripheral neuropathy is different than multiple sclerosis.

5:44
For example, it is not multiple sclerosis and people with peripheral nerve disease by and large don't have a problem from the process that's causing the peripheral nerve disease to affect the brain or the spinal cord. There are usually separate causes.

6:04
And so when we had talk about C I D P, or chronic inflammatory demyelinating, Pauline neuropathy, well, what is that exactly?

6:17
Well, if we go through the, the words themselves, the word kronick in this setting means more than a couple of months. So, people who have ...

6:32
have had symptoms that have been progressing over a couple of months.

6:39
And there is no, no specific blood test what we call a biomarker.

6:49
There's no specific blood test that tells you a person has the IDP or not, so you have kind of have to figure it out based on a lot of different kinds of information. And I'll get to that in a minute.

7:04
So the word chronic means more than two months, and that distinguishes ... from ... Syndrome, for example, which is that the other name for it is a IDP, which is acute, inflammatory demyelinating, Pauline our empathy end.

7:20
Acute means something that's happened over minutes, hours, days, or a few weeks, but no more than that.

7:30
So it's so a IDP or guillain Barre Syndrome.

7:35
People who have had a numbness or tingling and weakness that started and then stop progressing.

7:44
So in other words, you could still have many months later symptoms of numbness and tingling, and pain and paralysis, but you, but you're not getting worse all the time. In fact, you might even be getting better.

7:58
And that's guillain barre syndrome.

8:00
So, while it looks a little bit like ..., and in the beginning, a person can come down with sudden onset. So, in other words, I, you know, I'm perfectly well going along my business, and then one morning, I wake up and I have numbness or tingling in my hands and feet.

8:19
And by that, by the end of the week, I can't walk.

8:22
Um, most of the time, that's going to be a IDP or ...

8:26
syndrome, but it could also be see IDP because nothing in the definition that we'll get to says, how fast the symptoms are supposed to start.

8:37
But what the difference is, is that it's supposed to go on for months, and get worse over months, to B, C I D, P.

8:49
So that's the word, kronick, and that's how it's defined.

8:53
Now, inflammatory means that if you were to take a piece of this, of a nerve, and look at it under the microscope, you would see certain kinds of inflammatory cells, cells that are responding to something that shouldn't be in the nerve.

9:11
And that's what an inflammatory process is, um.

9:16

Millimeter, Excuse me.

9:19
So, um, inflammatory it is a marker that the immune system has gone awry, and see, IDP is a a condition that we call auto immune.

9:37
It means that your immune system has gone awry, and has attacked itself, has attacked its own nerves. And I often tell the story, and if there's anybody in the audience was my patient I probably told you this story before about the the guards in the castle story.

9:55
So my guards in the castle story is that if you think of your body as your castle, and you think of the guards of the castle as your immune system, you know that the guard's tend to know and distinguish who is a stranger coming into the body or into the castle and who lives in the castle.

10:19
And so they know to attack the stranger that's coming into the castle by, and then they know not to attack the people who live there.

10:29
However, when there is one of the guards of, you know, have had too much to drink, or their eyeglasses have gone awry or something. They get confused. And so they don't know who is living in the castle anymore and who's a stranger, and so they just attack wildly and without consideration of who they should be attacking or not.

10:54
So that's my guards and the castle's story.

10:56
And that is a way of explaining the very complex processes that are happening in the body, and people will see IDP.

11:08
It is an autoimmune attack attack against certain components of peripheral nerve and there are many other autoimmune disorders. Lupus, you may have heard of thyroid disease, very common, autoimmune disease, my senior gravis and so forth.

11:25
So, all of these are disorders where the immune system has gone awry, for reasons that we don't really understand completely and that starts to attack different parts of the body.

11:40
So that's the inflammatory part of the, uh, of the of the title of the of the disease. A poly neuropathy just means that many nerves, not just one nerve that's affected.

11:56
And you can distinguish appalling neuropathy from mono neuropathy for example, and the a manon neuropathy is would be something like Carpal Tunnel Syndrome, where a single nerve, the median nerve, it's the one that goes to these three fingers.

12:13
Makes them naaman might even make your thumb movement week.

12:20
And so, yeah, Poly neuropathy very often affect too many nerves to name and typically it would be it affects the feet first, but not necessarily and not necessarily primarily the feet are only the feet, but it can also affect the hands and when a poly neuropathy is advancing or progressing, and very often it will started the tiptoes and that's the reason is that nerves, that start in the, in the low back, because that's where their mother cells are, their origins in the low back.

13:05
And those law, nerves are very long. They go all the way down from your low back to your toes.

13:10
And so, if you sort of think about the different parts of your body, you can imagine that these are really long nerves, that require a lot of infrastructure to maintain their integrity and their health.

13:26
And so, and so very often and many neuropathy is like diabetic neuropathy, for example.

13:32
The toes become numb first, but unlike other month, like unlike most other neuropathy, ... causes a weakness as well.

13:43
So a diabetic, for example, who has a diabetic neuropathy because of look very long standing, maybe poorly controlled diabetes, will get numbness or tingling in their feet, which can be very painful.

13:58
But they don't typically have a lot of weakness which it helps to distinguish a C IDP from diabetic neuropathy.

14:10
Um.

14:11
Additionally, the weakness of the IDP is affects not only the feet, muscles are the muscles in the feet and the muscles in the hands, but also the muscles in the what we call the proximal limb, the proximal portion of the limb.

14:31
Meaning the portion of the limb that's closest to the center of your body. That's proximal. And this is distal. Meaning the hands and and so not only are the upper extremity proximal muscles affected but also the lower extremity small. Muscles are affected in C slash EBP making it difficult for people to get up out of a chair, to get up off the toilet seat, to get out of a car, To go up, steps to go down steps. So, not only do they have trouble walking on their tiptoes, but they also may have trouble doing these other things.

15:09
There are not too many other neuropathy, is that do that guillain barre syndrome. Of course, will do that as well.

15:17

And then there are a couple of other rarer ones and sometimes confused with CI. DP, and that's amyloidosis, and I don't want to spend a whole lot of time on amyloidosis, but I will say that those two diagnoses are often confused.

15:35
So if a patient has been diagnosed with C I D P and, and they, uh, get standard treatment, inappropriate doses and they're not getting better, you have to think of well, they either have ... that what we call refractory to therapy.

15:56
Meaning that it just, they have a particular kind of ... that just doesn't respond to therapy the way. Other ...

16:06
patients respond or they have a different diagnosis.

16:11
And sometimes it's hard to make that distinction very early in the, in the disease. But these two diagnoses should not be confused.

16:21
And they, it is an important consideration, and important to consider a different diagnosis because of course, it will be treated in a different way.

16:32
Then CIP.

16:35
So, I talked a little bit about the poly neuropathy. And then the last part, C I D P.

16:41
The D myelination, I should have said that before that P because it's C I D, the ... poly neuropathy but I need you to understand what a, what a neuropathy is before I can talk about the myelination.

16:55
So, what, does that mean, the myelination?

17:00
When we think of a nerve, you can think of a wire and very much like the wire on my desk. Here I'll put it up here for you. You can see that it has the plastic on the outside and then the copper on the inside.

17:16
Encryption nerves are very much like that wire in of course, much more sophisticated way.

17:23
Yeah.

17:24
The inside of the peripheral nerve is called the axon and the outside the plastic if you will of the peripheral nerve is the myelin myelin is the coating on the nerve that makes the nerve conducts electricity very quickly.

17:43

And, and the reason we need to have nerves, that conduct very quickly.

17:47
Because you need to send information very rapidly from your feet, in particular, to your brain.

17:57
The reason is, that's how you maintain your stability.

18:01
If you can't, if your brain doesn't know where your feet are, it doesn't know which muscles to activate to maintain your stability and to maintain your posture in an erect way. So then you start the wobble around maybe even topple over.

18:18
Which we call that the wobble we call an a taxi or a gate a taxi, and you don't have to worry too much about the lingo and the jargon here, but you will probably hear people say these words and so it's always good to be familiar with them.

18:35
And so, de myelination is the process where that inflammatory process that I told you where the guards are getting confused the immune system is starting to attack the myelin And and so, when the myelin the integrity of the myelin is disrupted then, the myelin doesn't work properly. And it doesn't send the signals back and forth across the nerve as quickly as it needs to.

19:03
And so we developed either paralysis, weakness, numbness, tingling, or unsteadiness, which again is an important feature of the IDP that sometimes is overlooked.

19:19
And so.

19:24
That sort of sums up the title of this, of this disease, are the, the, the name of it, if you will.

19:35
So, now, do we go on to make a diagnosis of ...? As I said earlier, there are no biomarkers, you know, when you have a pneumonia, you can get blood cultures.

19:48
And you can isolate the organism. When you have thyroid disease, you can measure the thyroid antibodies and the level of the thyroid hormone.

20:00
And when you have my senior gravis, you can measure the certain antibodies when you have coven.

20:05
You can measure antibodies, and that tells you that that the disease it has affected this person, There's no such thing.

20:19
Excuse me, There's no such thing. Or, Sea IDP?

20:24
So we have to rely on our are a few things.

20:30
Number one, we rely on the pattern, and as I told you, that this has to be a pattern of a progressive neuropathy.

20:38
So the patient has to give you a history of symptoms of a neuropathy that has been going on for a couple of months.

20:48
Number one, that's number one, and then those symptoms, they can be all pure sensory. So just numbness, tingling, and so on. So that's one form of ...

21:00
called sensory ..., very rare.

21:05
More commonly they give you symptoms of weakness.

21:07
I'm having trouble getting out of a chair, I'm having trouble getting off the toilet seat out of a car, going up and down steps and I have numbness or tingling in my hands and in my feet.

21:19
And so, that history would be consistent with a peripheral neuropathy but then you need to sort of dig into that a little bit more and refine that.

21:32
And so, you have to examine the patient, and there are certain features on an examination that the neurologist will look for, too.

21:45
Identify findings that are supportive of a peripheral neuropathy. You can't look at a patient and say, oh, yeah, clearly that patient SCID but you can't do that.

21:56
You need you need a lot of other information, so it's not a bedside diagnosis that you can make.

22:04
And so besides the clinical examination where people lose their reflexes, not always, but usually, they have weakness in certain muscle cells, like the ones I talked about, you know, the muscles of the feet, the muscles of the hands, the muscles of the upper arms and muscles of the legs and thighs.

22:28
You can't have bowel and bladder symptoms because then that doesn't quite fit with a diagnosis of .... You know, of course, you can have the IDP and problems with your bowel and bladder because you could have two different things going on.

22:43
But CIP typically does not cause disturbance in what we call sphincter function. So you should not have incontinence of urine or bowel as part of the illness itself or due to the illness itself.

23:01
And so So you have the clinical examination And then the next most important test is the EMG.

23:10
Now the EMG is the NRF test and it has two components. But it's called confusingly EMG Only.

23:19
It has the nerve conduction studies portion of the test and then the EMG part of which is that the needle examination or in our laboratory we call it a pin. We don't call it needles because pins don't hurt as much as needles and it shouldn't be that painful. But it is uncomfortable, certainly, and, no, no trip to the beach. And so a nerve conduction test is done on a particular set of nerves.

23:46
And sometimes you have to check a lot of nerves because not every nerve is going to be affected.

23:52
And what you're looking for is that the myelination that I talked about earlier where the the myelin is affected and that's really one of the only two ways of determining whether a nerve has the myelination or not.

24:07
one is by EMG. Another one's. Just take a nerve and look at it under a microscope. A nerve biopsy, which we don't typically tend to do except where an unusual circumstances. So most of the time you don't need a nerve biopsy, sometimes you do.

24:24
But most of the time, you don't.

24:25
and you can look at the nerves study and if it was done.

24:35
According to a particular protocol and a particular way, you can determine that this patient has de myelination or features where the myelin and not the axons are disrupted.

24:49
And as I said, the myelin is oh!

25:01
The myelin is the part of the nerve.

25:04
That helps to conduct electricity, and that's the part that's affected in the IDP.

25:10
The, um, the the axon is what is typically affected in people who have a diabetic neuropathy for example and that's how we can make the distinction between diabetes and ..., excuse me.

25:28
Yeah.

25:31
OK, I'm getting over a cold and I have a bit of a tickle in my throat.

25:41
So The nerve conduction test is interpreted according to some guidelines and to help, as I said the diagnosis is a difficult diagnosis to make.

25:55
So a group of experts have gotten together and back in 2000.5 11 and made a set of diagnostic criteria, as we call them. And a diagnostic criteria is just a list of things that a patient should have.

26:17
In terms of history, examination findings, EMG, findings. And so, there are features of the myelination and those criteria, you can actually look them up.

26:30
They're called the European Academy of Neurology, Peripheral Nerve Society Criteria, and those were originally published in 2010 and then revised in 20 21.

26:43
And, again, these have been published, and, and they are what we call fairly sensitive, and specific for the disease.

26:52
Sensitive means that if people, you take 100 people with the disease, a large number of them are going to have are going to meet those criteria, not everybody.

27:05
So, there are, some people will see IDP, who don't fit all of those criteria, and that always makes it a challenge. And that's why I think it's always good to be seen if you have a diagnosis of .... Especially if the diagnosis is not entirely clear by someone who sees the disease fairly often.

27:27
And you can find people like that in an Centers of Excellence for ... syndrome.

27:37
And I'm sure the foundation for peripheral neuropathy can can guide you in that regard. Somebody who's close to your home, or some center that's close to your home, and it's always good. Sometimes it's good to get a second opinion, just for confirmation of the diagnosis.

28:01
And so using those criteria, which, as I said, are fairly sensitive for the disease and excluding certain other conditions. So, you know, there are other diseases that look like .... I said, At the top of the hour, I talked about amyloidosis.

28:23
But there are a number of other things that can, that have to be excluded, that you have to make sure that the patient doesn't have, before you make a diagnosis of a definite SCID P or ..., and then.

28:40
And then you go on from there.

28:43
Another, another diagnostic modality, if you will, is a spinal tap.

28:49
So you don't need it in every case, depending on the situation, but it does give you helpful information.

28:57
And, like and guillain barre Syndrome. where the spinal tap will have very few white blood cells in it. Those are the inflammatory cells. And even though this is an inflammatory disease, as I said, it's not an inflammatory disease of the brain and spinal cord. So you really shouldn't see a lot of inflammation, or inflammatory cells, or white cells in the spinal fluid.

29:22
But you should see elevation of the protein, So there are criteria and there are numbers that haven't been published and people accept as being abnormally high.

29:34
And it would support a diagnosis of ..., in addition to the spinal tap.

29:42
You can get an MRI, for example, an MRI is a very nice test set looking at the soft tissues around the spinal cord and brain, and it can look at the nerves as well.

29:58
And as spine MRIs are now becoming very sophisticated.

30:03
And one of the criteria that we can rely on for the diagnosis of ...

30:11
is peripheral nerves, that enhanced when you give got a linear enhanced, meaning they light up on certain sequences of the MRI.

30:23
And so, using the MRI as a simple, or as supportive evidence of peripheral nerve the myelination, when the nerve, um, is inflamed and disrupted, the structural integrity of the nerve is disrupted. There is a breakdown in the barrier. So ordinarily, there's a wall, if you will.

30:51
That prevents the blood elements from getting into the nerve so-called the blood nerve barrier.

31:00
When that barrier is disturbed, because the nerve is inflamed, and you give the patient gadolinium that gadolinium is going to seep into the nerve from the blood, You'd give the gadolinium by injection in the blood in the, in the vein. And it circulates in the body.

31:17
And then that gadolinium will deposit in the nerve, and you can see it on MRI.

31:26
So, that also is a supportive criteria that is accepted if for the, to support the diagnosis of ....

31:36
Um, finally, you win.

31:41
You can't make a diagnosis. Or you are considering something else like amyloidosis or vasculitis affecting the peripheral nerve.

31:52
You can take a biopsy.

31:53
And typically, we biopsies are old nerves, so a sterile nerve is if you look at your foot on the ankle on the outside of the foot.

32:04
Behind it is a little, tiny wire.

32:09
And I'd show you my foot, but that won't look nice. So I'm not going to do that.

32:14
So there's a nerve that goes right behind the ankle and you can actually cut that nerve out.

32:22
We don't like to do that, too, too frequently. The reason is that it can cause pain in the area of the nerve.

32:30
It can cause some extra numbness when you've taken the nerve away and And sometimes you don't make a diagnosis even when you do when you do a nerve biopsy. So it's not the kind of thing that we enter into lightly and like to really reserved for cases where not only were not thinking about the IDP, but we're thinking the patient may have something else.

32:57
So, those are the pretty much the diagnostic modalities that we can use to make the diagnosis of CIT.

33:08
So, once you've established a diagnosis, now you want to get into the treatment of ... and the treatment can be complex and it has to be individualized.

33:19
No two ... patients look alike or treated the same way, or for the same duration of therapy or anything like that.

33:32
So you really have to tailor the therapy to the patient's needs.

33:38
And the goal of the therapy is to improve quality of life.

33:45
That's the goal of any therapy, of course. See, IDP in and of itself, is not a fatal disorder.

33:51
So it's not going to, it's not going to kill you, but obviously, it will cause severe disruption in the quality of your life and, and that is the goal of therapy.

34:07
So I'll start with the, um, sensory symptoms, which may be significantly problematic. And then I'll talk about the motor symptoms.

34:19

So the sensory symptoms are the numbness, tingling, pain, Burning that people complain about very often as people will say that they feel like they have pebbles in their shoes or that their socks are bunched up or that the or the socks are too tight around the foot.

34:41
That's those are very common burning like I'm walking on sand on hot sand, Those are symptoms of many peripheral neuropathy but in particular also CI when you have symptoms like that Unfortunately you can never really get rid of the numbness and the tingling per se but you couldn't get rid of the pain associated with the numbness in the things.

35:11
And you may not maybe get rid of as too strong a word.

35:14
Maybe take the edge off of it so that you are comfortable or that the quality of your life has improved because you can take the edge off of the pain.

35:28
It is well known that people with chronic pain can be very productive.

35:34
And so, just having pain alone doesn't mean you can't be productive, but, obviously, having pain, you can't be as productive as you would want to be if you are pain free.

35:45
Um, and, and, and, nowadays, as we move away and have moved away from the use of narcotics, which, we used to use more often in the treatment of the IDP.

35:58
And I can't really remember the last time I gave somebody narcotics for ..., You can use a lot of the treatments that are available that are non narcotic non addictive. And you can always throw them away without going into withdrawal.

36:16
And without having the question of, Well, it's the patient, um, having relentless pain because they have an addiction for the narcotic. Or is it just you're not able to control the pain?

36:33
There are occasions when people will need narcotics and I think a way and so it's not just to dismiss the use of narcotics out of hand.

36:43
It is, too, temporize, they use a use of narcotics and now that big, because, because of the dangers that we recognize narcotics to have, we have four, I prefer that the narcotic administration be given by a pain specialist, These are people who are trained in the use of narcotics. They know how to administer them, how to withdraw them, when it is appropriate to escalate the dose, and when it's appropriate to reduce the dose.

37:20
So when I have a patient who's just not handling their pain at all with my therapy, I then refer them to a pain specialist, and the drugs that I use are very common drugs.

37:36

So I use gabapentin, I use pregabalin lyrica gabapentin, it's also called Neurontin, I use topamax the pyramid, I used depakote, I use, nor Trip, the Lean, which is also called Pamela. These are all of these are old drugs. They're pretty harmless in terms of serious side effects. Like like any drug, they all have side effects but the side effects are usually fairly well tolerated.

38:06
And the, and acceptable.

38:13
And as I said, if if the mie therapy is inadequate, then I refer the patient to pain management, and we co manage the patient's pain.

38:25
And then, um, the, because the other feature of the IDP and the really, in addition to the pain and numbness and tingling, or the motor symptoms, that condition now needs specific therapy.

38:44
And this is where the immunotherapy is, where you try to calm those guards down and tell them like, chill a little bit and stop attacking the body, stop attacking the nerves.

38:59
And, and and the commonly used drugs, are immunoglobulin.

39:05
So, IVIG, ..., and Prednisone, those are very commonly used very well established.

39:14
Good studies with placebo controls, showing that you can control and improve patients, weakness.

39:28
Um, so how are these drugs given and, and what is the, the problem with them?

39:38
The steroids can be given either as a friend his own tail, which is what I prefer or they can be given as even pulse steroids over periods of time in big doses, Recurrently over time.

39:56
The problem with ... Zone or any steroid given in the high dose. For a long enough period of time is that they are a double edged sword.

40:08
They will improve symptoms but then they all cause other problems on the other hand, and so they have to be used judiciously and over time you can't really use a high dose. What is a high dose?

40:27
Uh, a milligram per kilogram, so if you away, if you weigh about £150, you are about 70 kilogram person somewhere around there.

40:41
And so, um, a milligram per kilogram so 70 milligrams, 80 milligrams, 100 milligrams a day is a high dose of prednisone.

40:56

I don't like to use those high doses for more than a month or two at the very most. Just to demonstrate that the patient does respond or does not respond.

41:07
Um, the nice thing about Prednisone is a very cheap.

41:12
Just they, know, a few cents a month or a couple of dollars a month, or get, your pilot Prednisone.

41:20
So the probably the cheapest drug going except maybe gabapentin and those other drugs are also pretty cheap because they're generic now.

41:31
Um, the side effects of Fred in his own are are numerous and you have to be careful with them. They can cause cataracts, for example, pregnancy causes diabetes. So if you have a diabetic, who has ..., and there is information to suggest that people with diabetes are more prone to the inflammation, to the inflammatory process, that results in the IDP.

41:59
Yeah.

42:00
You have to be careful using prednisone.

42:03
They also make bones thinner.

42:06
So if you already have osteoporosis or osteopenia, they will leach the calcium out of the bones and make the bones more brittle prone to cracking.

42:17
Even spontaneous cracking.

42:19
So you have to be careful with steroids that can raise the blood pressure. They make wound healing very poor so the skin won't heal very well when it's injured.

42:32
So these are the problems with with Prednisone used in high doses. It's wonderful because it makes people stronger very quickly.

42:41
It also causes mood irritability. So you have to be, you have to be careful about that.

42:48
Immunoglobulin is the other well accepted.

42:54
First or second line drug that that we use and the FDA has approved a sub Q immunoglobulin.

43:04

There's a whole bunch of them on the market right now, CSL bearing makes the high zandra, there are others that are equally effective.

43:14
Probably, although, none of them have been compared, head to head.

43:18
But anyway.

43:21
The drugs that are used some Q under the skin you can give them to yourself or somebody can give them to you, but they, you need to have an IV loading dose first or if you can't tolerate the IV for whatever reason.

43:41
You know, people with IV get sometimes will get a headache from the IV.

43:45
Or, they get nauseous from the IV, or they don't have venus' access, and they would need a port.

43:51
So to be able to access their veins.

43:54
And then or they travel for work and they can't get IV immunoglobulin, The nice thing about immunoglobulin compared to other medicines except bread and zone is that it's not as expensive as the newer ones, and I'm going to talk about some new ones in that in a second.

44:16
So, but it's still an expensive drug. And it has to be approved by the insurance company.

44:24
And you either have to go to an infusion sweet to get it, or you can have home infusion, which makes it very convenient for some people.

44:33
Aye.

44:34
IV, IG or sub ... are pretty safe drugs. These drugs have been around since the fifties. They are used for other diseases as well. Most commonly for common variable immune deficiency where people don't make enough IG, we all make a little bit of IG.

44:53
But you need a certain amount to prevent infections, for example.

44:57
So, and IVIG is not really an immunosuppressant, the way prentiss zone is. It is really an immune modulator so it alters the immune system. It tweaks those guards that I talked about a little bit.

45:15
In terms of safety, like I said, that they're pretty safe, to be given at home, it, there are risks associated with them, and, of course, you have to talk to your doctor to make sure that, you

know, if you have a heart condition, for example, it may not be appropriate for you. If you have very high blood pressure.

45:35
That is not well controlled, it may be inappropriate for you. So, if you have to really talk to you your own doctorate to have tailored therapy, as I, as I mentioned.

45:49
And, um, and lastly, recently the this is not yet FDA approved, but there are new drugs now the so-called ... inhibitors that are the first time that a new medicine for.

46:09
See IDP has been come onto the market.

46:16
The, uh, drug is not that. The top line results were just published, But they have not been FDA approved. And they really haven't been published for in peer reviewed journals yet, to really look at the data.

46:34
and and make sure that it's all COPPA set, but the company says, it looks like it's going to be an effective drug. And so I'm excited that we're going to have more options for patients.

46:48
So.

46:51
I think, you know, the, the ebb overall, I, I like treating this disease because I like to help people, and this is one way that I can, I can help people.

47:04
I think you can get a lot of benefit from treatment of this disease, assuming that the diagnosis has been made correctly, that you're getting the medicines that you need to get in the appropriate doses through the appropriate means and that, you're being monitored by people who have an idea of what they're looking for.

47:35
And lastly, I would urge you all, because we really only have a few drugs, there are a few others that I didn't mention methotrexate, cyclo sporran, And the others sell Ceph where the data is kind of iffy, whether they really work, or not.

47:56
And they may not work for everybody, they may work for some of them, for some people, but there are certainly not as good as the prednisone and and the immunoglobulin that image.

48:09
And, then, the last plead that I would make to you before I wrap this up, is, is to participate in clinical trials. And we, if you go to a big academic center.

48:25
There are a number of companies now that are doing trials at NCI D P with both.

48:34
Yeah, F CRN inhibitors, which are those, that new drug, a class of drugs that I talked about.

48:43
And so clinical trials, you're not a guinea pig.

48:48
You're being offered a therapy that is not commercially available.

48:53
And by and large, you may be in the placebo arm of the, of the trial, for a short period of time.

49:07
But then you're being offered a drug, that you can't get, that your insurance company and your doctor can get for you, which may be effective, and maybe even more effective than the current therapy that you're getting. So clinical trials are a good thing.

49:24
They don't think that they are.

49:28
There's any, you know, sometimes people are understandably skeptical that, you know, I don't wanna be a guinea pig, and, but you're not being guinea pig.

49:38
I think if you're not completely satisfied with the therapy that you're getting, it is important to participate, to advance the knowledge that we all have in trying to understand this disease, and that's what clinical trials, in part do.

49:57
Um, so, I think I'm going to stop here, Lyndsey, and if there are any questions, I'd be happy to entertain them.

50:11
Yes, hi, thank you so much, doctor Kahler, this was amazing. There are several questions that are coming in so I'm gonna try to ask as many as we can in the remaining 10 minutes in our session.

50:26
So, you had mentioned a lot about other forms similar to ... including GBS. Is there any similarity or relationship with Mac anti mag neuropathy? Could you speak a little bit to that? There were several folks that were curious about that relationship, if if there is any.

50:46
Yes.

50:48
So anti magas mag is myelin associated glycoproteins and that is A part of the peripheral nerve.

50:58
And sometimes, patients will have a group of cells that are making this antibody.

51:09
It does have some features similar to ..., but that neuropathy is really treated by the hematologists.

51:17

So, this is where a multi-disciplinary approach is required, where you need. You need a hematologists or him on a person to participate, because very often, anti mag, while it can respond to IVIG Sometimes, is treated with her talk, san or other or other drugs.

51:39
So, it is part of the workup when you first evaluate a patient with .... You look for these mag antibodies as well.

51:54
Great, thank you.

51:55
And could you explain a little bit more about the cause of Muscles Wasting, why that continues to happen? And if there is, if, if we do know that answer, at least?

52:09
Sure. So mussels waste for two, for two reasons.

52:14
Number one, there's what we call this use. And Waste Muscle Wasting, the, the, the, the technical word for it is atrophy.

52:24
So muscle atrophy happens when you don't use a muscle.

52:29
So if any of you have been in a cast for a few, even a few weeks or if you just sit in bed for a week, this is why you shouldn't work from bed. And you gotta, you gotta get up and walk around.

52:44
If you sit in bed for a week, and it's actually been shown that your muscles shrink, they become a trophy after one week. So if you've been in a cast for, for even a week or a couple of weeks, you'll notice that your limb is really skinny. That's disuse atrophy.

53:01
Then the other reason why muscles atrophy is if you cut the nerve to them or injure the nerve that's going to the muscle. The nerve is connected to the muscle, and they have a very intimate relationship with one another. They feed each other. And so, when the nerve is damaged, the muscle is heartbroken and shrinks.

53:26
So, you can see, for example, people are very bad Carpal Tunnel Syndrome. They'll have a thinning of the what, we call the ... Eminence or the meaty part of the of the palm.

53:39
And that is atrophy. because that nerve has been injured.

53:45
And is it common for treatments to use any or solid acid or anything like that?

53:53
With, with C IDP or other forms of neuropathy is I'm sorry what kind of acid?

53:59

one, so one of our, one of our constituents is curious if you know anything about ... acid.

54:07
So there are a number of, of over the counter, um, uh, hayles, concoctions, at the end.

54:22
Some of them may be affected in alleviating the pain. They don't really I've never heard of anything that works for the weakness, but alleviating the pain of the numbness and tingling and the burning.

54:40
There are many, many different things that people have, have used and have told me that works for them.

54:46
The thing is that none of these things have been studied in a scientific way, you know, using a certain population of people comparing it to a sugar pill. So I can't really say, Oh, categorically. this works. Or this doesn't work, or this works in 60% of people. I can't really say that at all.

55:09
I don't discourage people from using things that they like to use and they feel good on.

55:14
I would just say, like, don't spend a lot of money and, and make sure you're not taking something that's poisonous and certainly not injurious to your nerve because, you know, vitamin B six, for example, can be toxic to nerve.

55:28
So you don't want to take too much of something that can injure your nerve.

55:33
But I think, you know, if it makes you feel better and you don't mind spending the money out of pocket because then insurance companies usually don't pay for those things.

55:40
I say, Go for it, OK, and you mentioned briefly about vitamins, obviously. It's always good to test to see what your deficiencies are.

55:50
So you don't just take things because that's not a good idea either, but do you have any other comments about diet as well specific to see at EP if there's anything that helps to improve symptoms with respect to what you can consume?

56:07
Yeah, uh, I don't think that there are particular foods that that help or hinder patient with CI ADP. What I would say is that you need a varied diet.

56:24
So it's very important to eat from all the food groups.

56:28
You know, vegetables, meats, fish, chicken, because you need all of those components in food, to make sure that your nerves remain healthy. So, a varied diet is all, I would say.

56:44
Some people just don't like certain things, and that's fine. But there's nothing in particular, I would say, that you should eat or avoid.

56:55
Makes sense.

56:56
And then, the last question that I'm going to try to ask before we run out of time, is there any relationship with the IDP and sjogren's syndrome?

57:09
So, CI DP and sjogren's syndrome are both autoimmune disorders.

57:14
And so, it's not uncommon that people can have, it's not common that people have both because they're both kind of rare diseases. But it's not uncommon for sjogren's patients to have a neuropathy.

57:27
Now that neuropathy is not see IDP, but they do get a neuropathy for sure.

57:32
And it can be as sometimes, it can be a very bad neuropathy.

57:37
but the only relationship that I know of is that they're both autoimmune disorders and when you're prone to one autoimmune disorder, are often prone to another.

57:48
Just whatever it is, that causes autoimmunity, OK.

57:54
Well, great, well, this is extremely helpful, doctor Carla. As I mentioned, we had a lot of other questions that are coming in. We'll make note of those. We'll try our best to.

58:05
We'll follow up following this presentation. Just to try to address some of those questions, that we haven't been able to today. So, for those of you that weren't able to get those addressed today, I do apologize, but we'll try our best to follow up at the, at a later date.

58:21
And, at this time, I just want to thank you once again, for presenting today's lending your expertise on this important topic. On a special thanks to our sponsors, that sponsorship program of CSL bearing. Once again. And for those that are watching, as always, thank you so much, for joining us, for being a part of our work. We hope you liked this webinar, We will be sending out a survey following. So please do share some feedback, this is how we continue to improve our programs. And please continue to support us as much as you're able to every dollar matters. And that allows us to keep doing things like that. So, again, thank you so much, everybody. Doctor Hal it's always been our pleasure, and we hope that you guys all have a good rest of your day. Thank you.

59:09
The vow.