Transcription

This transcription has been lightly edited to enhance readability by removing repetitions and speech fillers.

00:00:05 Beaudin

Hello.

I am pleased to be here. I'm Marie Beaudin. I'm specialized in neuromuscular diseases. I'm in Quebec City, at Hôpital d'Enfant Jésus where I did my training in neurology, after which I completed my specialty in neuromuscular medicine at Stanford University, where I worked on autoimmune diseases. So that's what I did. I have a clinical interest and a research interest on risk factors and treatments.

00:00:50 Picher Martel

Hello, my name is Vincent Picher-Martel, I'm a neurologist specialized in neuromuscular diseases at CHU, Quebec City. I started my practice in July and as Dr. Beaudin. I was trained in Quebec City and I trained at Harvard for two years for my specialization in neuromuscular diseases, and I am also a researcher and a clinician here in Quebec City. I focus on research, muscular dystrophies, and I also see a lot of neuromuscular patients in my practice.

00:01:39 Grant

And I'll now invite our other panelists to introduce themselves, and then we can begin the Q&A. So we can go with Dr. Beadon and then Dr. Vasjar.

00:01:50 Beadon

Hi there. Nice to be here. Thanks for the invite. I'm Katie Beadon. I am a neuromuscular neurologist in Vancouver, BC. I did part of my training here in BC and then a fellowship at the Pitié Salpètrière Hospital Neuromuscular Disease Centre in Paris, France. I'm looking forward to hearing your questions and answering them as best I can.

00:02:08 Vasjar

Good afternoon, everybody. I'm a pediatric neurologist, neuromuscular specialist at the hospital for Sick Children in Toronto, and I have looked after the pediatric CIDP and GBS patients for the last 35 or 40 years maybe, and at any given time, with my colleagues at Sick Kids, we have about 10 to 15 CIDP patients, so I'll be happy to answer any questions which relates to pediatric CIDP patients.

00:02:47 Grant

Well, thank you all for introducing yourselves. We're all very eager to hear your answers and to hear the interesting discussion that we're going to have. So we'll start with a few questions in English and we'll alternate back and forth between English and French. So feel free for the participants to ask your questions in whichever language you prefer.

00:03:08 Grant

Our first question is for Dr. Beadon.

As a longtime participant in support group meetings, there seems to be an increasing amount of patients who are initially diagnosed with GBS being re-diagnosed with CIDP. So there's two kind of questions. The first question is, are neurologists seeing an increase and were they always CIDP?

00:03:36 Beadon

That's a really good question. So what we refer to is sub-acute onset of symptoms. So those people who fall into the time frame of symptom development in that 4-to-8-week category, we're never really sure if they're going to end up being a monophasic illness like GBS or if they're going to be a chronic illness such as CIDP and it's a bit of a pattern of practice dependence on where you are, whether you treat them for three months or just treat them once and see what happens. I think these patients have always existed and personally, in my practice, I haven't seen that there's been an increased incidence of people who initially I think are GBS and then end up having CIDP. That's something that we've sort of seen all along.

One thing…Sorry, I am at home with two children. You may have just heard a giant bang in the background. One thing that we are more aware of now, though, is these, these nodal-paranodal presentations and inflammatory neuropathy, and I think Dr. Katzberg spoke a little bit about that in this session yesterday, and we know that these patients can present in a more acute fashion, but then have a chronic illness associated with that. So not that it's necessarily increased over time, but I think we're getting a bit more of an understanding. So I think that was the first question. And then, sorry, was there a second part as well?

00:04:57 Grant

I think that answers the question about whether there's an increase that you're seeing or if, and also if patients who maybe started out with GBS, but then they turned out to be CIDP, that sort of monophasic versus chronic.

00:05:13 Beadon

Yeah, yeah. And it really is. It's sometimes difficult to predict. There are a few things that we think make us think it's more likely to become a chronic illness, but a lot of the time it is, unfortunately, a wait and see sort of approach.

00:05:27 Grant

Is there anything that you see in, let's say a GBS patient or presumed GBS patient that makes you think, maybe this patient doesn't actually have GBS, maybe it will turn out to be CIDP?

00:05:40 Beadon

Well, that's a good question. So, going in the opposite direction, I don't, I don't think so. We know that patients who end up having the more chronic version are less likely to have things like cranial nerve involvement. So weakness in the face or difficulty with swallowing or difficulty with breathing, but minor or sorry, more mild versions of Guillain-Barré can also present without those symptoms.

So I would say, if there's a patient who was progressing on the slower end, because a lot of our GBS patients, it's a day-by-day change in their symptoms. But if we're hearing that story that it's more of a one week to the next week that they're seeing those changes and they don't have any cranial nerve involvement, then that might make me slightly more suspicious, but it is often difficult to tell at the beginning for sure.

00:06:33 Grant

Thank you. Our next question is for Dr. Vasjar. Is subcutaneous immunoglobulin being used at Kids?

00:06:43 Vasjar

It's not easy to obtain, really, access to home care for pediatric patients to receive the subcutaneous IVIG, we have done it only in the one case so far and it wasn't a patient with CIDP.

We used the organization called OnePath, which is a cross-Canadian organization and so it can be organized. It's not the easiest way to do it, you know. For example, in the adult world, it's much easier to access the home care, you know, healthcare, healthcare providers, but it can be done. So we don't do it too often. We have done it on one kid so far.

00:07:40 Grant

Thank you. I'll ask a question in French now. It's a bit of a longer question that I'm going to read.

I was prescribed a high dose of gabapentin for pain during my GBS recovery after years of being on it. I asked my family physician whether I needed it anymore. And they didn't want to handle dosing or weaning me off from that medication, because it wasn't that doctor that prescribed it to me. So what can I do? How can I know that nerve pain medication is no longer necessary or can be reduced?

00:08:22 Picher Martel

Thank you for the question. So pain medication? Yes, eventually you can try to be weaned off pain medication. Generally it depends on clinical symptoms. When there's no more pain that's present. So generally, we have to wait a few months, or a year or two, before beginning the weaning process. If your doctor is not comfortable with that, there are other possibilities. If you have access to the neurologist that treated you recently, that's an option.

In my practice, I think that pharmacists are really helpful in that respect. They have some freedom in terms of weaning off someone from the medication, or at least lowering the dose. So that's a possibility with a pharmacist who can help you over a few months and sometimes, if you have a doctor who specializes in treating pain, or if it can help, there can be other doctors, like rehabilitation medicine physicians.

It's not necessary to keep taking pain medication if you do not have any more pain.

00:09:47 Grant

So are there contraindications in terms of lowering a dose or, or completely weaning off pain medication?

00:10:01 Picher Martel

No, I don't think so. There's no contraindication, medication like gabapentin and others, you have to go gradually with some medications, but you can actually wean off the medication eventually.

00:10:06 Grant

Thank you. Next question to Dr. Beaudin. Would you recommend acupuncture for pain? Are there other treatments that you would recommend? What about hypnosis as a potential treatment? Could this help with nerve regeneration? So there are two parts, pain and then nerve regeneration..

00:10:42 Beaudin

OK, very good question.In immune neuropathies like CIDP and GBS, we have to distinguish the core treatment for the disease with IG or plasmapheresis versus treatment for other symptoms, for example residual pain, weakness and other symptoms and presentations.

If for pain, for example, someone who has an optimal treatment for their condition, but still experiences disabling neuropathic pain has a number of therapeutic options. What I use the first, as a first line is drugs, and medications that have good evidence. So, for example gabapentin, duloxetine and, and so on. So there are different drug options, but patients can be interested in non-pharmacological options like acupuncture or hypnosis. So first of all, I would say that there is no specific evidence for GBS and CIDP, as far as acupuncture is concerned. We can perhaps extrapolate from literature on neuropathic pain in general, but there are limitations because the underlying mechanisms are different.

But let's take acupuncture first. If we look at neuropathic pain, there is some evidence for neuropathic pain linked to chemotherapy in cancer treatment, so for acupuncture or hypnosis, we do not have large studies with control groups so that we can control with the placebo effect. So we cannot warmly recommend them, but acupuncture, give[s] us some evidence. For example, neuropathic pain from chemotherapy. Some patients can see an improvement in their pain scores. Hypnosis, now, there's even less evidence. When I was trying to do some research

on that, there are very few dedicated studies with good cohort and good controls in terms of neuropathic pain.

A lot of patients are interested in these kinds of treatments. If you're interested in them, of course, you have to make sure it's safe. Generally acupuncture and hypnosis are safe options, and it shouldn't be too costly. If it's not too costly and doesn't put you in a difficult financial situation and you want to try them, then some patients can find some benefits with the limitations I've just mentioned.

We do not have a lot of studies, so it's not something that I would recommend in my practice, but it's not unreasonable for patients to try them.

Was there another part to your question?

00:14:18 Grant

Yeah, regarding treatments to encourage nerve regeneration.

00:14:18 Beaudin

So that's another chapter. We're coming back to the core treatment of the disease. Can acupuncture and hypnosis help someone to heal? In the literature that we have, there's no evidence that acupuncture and hypnosis can help heal nerves. What you want to do is treat the underlying condition. So I think acupuncture and hypnosis are more treatments for some symptoms or the management of pain. But they're not going to treat or heal nerves, and there's no evidence showing that it could regenerate the nerves.

00:14:52 Grant

So I've asked a round of questions to each of our panelists, and we want to make sure that we're having discussions around the questions. So even though I'm directing a question to one panelist, I invite you all to chime in if you feel like there's something you'd like to add, or if you want to have a discussion around a specific point, I think it would make for some really interesting discussions and maybe new questions might come up that way, so we'll switch back to some questions in English.

Dr. Beadon, is there validity in testing severe GBS cases for Contactin-1 or NF155? Would that knowledge alter the course of treatments for those patients?

00:15:39 Beadon

I think that's an excellent question and I think it's one that we don't have a complete answer for yet. The fact of the matter is that these tests take time, so even if they are available in your centre, they're not going to be available that day or the next day. So

we're going to have to treat the patient in front of us without that knowledge, at least in the short term.

The place where I see them to be the most useful is in those patients where we're not seeing them respond to therapy as we would like them to. And so, those patients who don't have any response to IVIG or specifically are continuing to decline beyond that first couple of days after we've given the IG and we're really worried about that patient or someone who's had, having a really prolonged course and not seeming to turn around a little bit.

In terms of, if it would change our treatment strategies, what we know is with the nodopathies, they are less likely to respond to immunoglobulins than those patients who do not have nodal or paranodal antibodies. That being said, many of them do still respond. And so even if we knew that they had an antibody present, they likely still would be treated with IVIG first because it's safe, because it's available. Because it's, we're able to give it pretty much right away. But if patients are not responding as we expect them to, and we are able to identify an antibody early, then at least for me, I would definitely move to one of our, I would escalate immunotherapy faster than I would for a patient that, if I didn't know that they had an antibody present. So, for instance, we'd move to the B-cell suppression therapies such as rituximab if we knew that they didn't respond to IVIG and had an antibody present.

The real challenge right now is in access to that testing. So in my province right now, you have to basically pull teeth in order to get them to pay for that testing, unfortunately.

Even though it's done at my centre and in other places, it's much easier to access, so I don't think we're going to see widespread testing of nodal and paranodal antibodies in the acute phase of GBS soon, but I think we're probably going to be using it more in the realm of CIDP and hopefully we'll get better access for those patients over time.

00:18:01 Grant

And this, this follow-up question, you know, it could be for anyone on the panel because we're all from different places in the country and things are often done differently depending on where you are. But how are you seeing these tests trending? Do you find that we're using them more and more?

Do you expect that we'll be using them for all the GBS patients or all the CIDP patients or do you want to reserve them for certain types of patients specifically?

00:18:37 Martel

Maybe I could, I could speak for Quebec, or at least in Quebec City, it's fairly easy to obtain a test for example, paranodal antibodies, but I agree with Dr. Beadon, I think that tests should be reserved to refractory cases where there's no good response to plasmapheresis or IG. And in my case, I would use it in the cases of CIDP, but for, and also for GBS patients who do not respond, or not as well, and generally we can see that and have a response or a result in about 3 or 4 weeks.

00:19:28 Grant

Thank you Dr. Picher Martel.

The next question is still related to antibodies and it's for Dr Vasjar. So in the pediatric world are there other cases of auto antibody variants in kids, and are you born with these auto- antibodies present?

00:19:46 Vasjar

I'll answer your second question first. Obviously, the antibodies are required as the child gets older and older, is exposed to different antigens and eventually develops these bad antibodies, which will fight to the peripheral nervous system, the nerves. So no, we are not born with these antibodies, so they are not genetically predetermined. And in the pediatric world, we haven't done any, nobody in the world has done any major testing on these NF155 and the antibodies. So I think it's still a way to go and it's a bit of a grey zone for the pediatric patients at this stage.

I don't know if anybody on the panel has had any children and tested for these antibodies, but we haven't done it yet at Sick Kids.

00:20:58 Grant

Yeah, it sounds like it's an area of research and still some questions there and there's always some challenges when there aren't that many patients that have these conditions to do these studies and investigations. So we'll definitely keep an eye out. Hopefully, these studies get done. So we have some answers regarding that.

Next question, for Dr. Picher Martel.

00:21:34 Grant

My question is as follows.

I have CIDP. I was stabilized on IG every two weeks. I'd like to know more about the plasmapheresis treatment. Is it plasma transfer and can it replace IG treatment? I feel good, but if there's a possibility of remission, I'm open to trying it.

00:22:04 Picher Martel

It’s a very good question. Plasmapheresis is really a plasma transfer, which the idea is to remove IG's or antibodies that we have in our blood [that] we believe attack the nerves and antibodies are in the plasma. So we replace the plasma with antibodies, we replace it with a plasma that often is like a protein solution. There's a plasma exchange that is done. We have no evidence that plasmapheresis is better than IG. We think both are equivalent. There have been studies that compared both therapies, and often both therapies had the same efficacy. So you wouldn't say that replacing IG with plasmapheresis could lead to remission.

In a patient where a plasmapheresis works well or IG works well, we tend to favour IG. It's easier because with plasmapheresis, you need a catheter, you need a kind of entry port and there are risks of infections and it's not easy, it's not comfortable for patients and you need more advanced facilities as well. Not all the hospitals do plasmapheresis, but IG is easy to administer everywhere. So to answer the question, I do not think that there is more hope in changing from IG to plasmapheresis in order to encourage remission. We move plasmapheresis, for people who do not respond well to IG, and in more acute cases of GBS.

00:24:06 Grant

Thank you. So with that type of patient that is stable on some treatment, for example IG every two weeks, after some time, do you try to change the therapy to see if there can be an even better response? Or do you think a patient that is stable should stay on this therapy as long as there's no deterioration and there's no change?

00:24:34 Martel

So it depends on the patient's history. I would say that in my practice, when we have IG patients every two weeks, we tend to, if they're really stable and the disease is controlled, we try to make IG administration less frequent. For example, every three weeks or every four weeks and see how it goes, or even six weeks in some cases with the stabilization with another medication, and it's sometimes possible to stop IG. We'll talk about it in another question later on.

It’s the same thing with plasmapheresis, where there can also be corticosteroid medication where you can lower the dose, which is the objective in some cases to lower the dose. So if it's a patient that's stable, we try to make the IG administration less frequent.

00:25:34 Grant

Now, Dr. Beaudin, if you've had GBS or CIDP, are you more susceptible to developing other autoimmune diseases?

00:25:37 Beaudin

That's another excellent question. Yes. Generally, in terms of autoimmune diseases, we see it frequently. There can be comorbidities, but that being said, the data that we have in terms of CIDP is not something that happens very often, compared to other autoimmune diseases. We have perhaps some conflicting data.

There was a Dutch study showing the percentage of patients who had an autoimmune disease that could, 9% could have another disease and then there was an Italian study which was actually questions, polling questions showing that there might be higher risk for patients to have a, then another autoimmune disease, but the disease that we see more frequently that's associated with CIDP is diabetes. We see it in a number of repeated studies, more patients with CIDP that also have diabetes. And there's also difficulty in a diagnostic, because there can be overlap between a different kind of neuropathy and CIDP and, so there are different issues. In order to see whether there is an association. So I would say, to conclude, that it is not necessarily the disease with which there's a lot of autoimmune comorbidities although we do not have a definite response.

00:27:47 Grant

Thank you. I hear a lot of patients talk about other immune dysfunction. Is that the case for people that have autoimmune diseases? Do they have a weaker immune system? Are they more prone to have other infectious diseases, or is that not the case?

00:28:10 Beaudin

That's a really broad question and it's quite complex.

There are some autoimmune diseases that can be associated with some dysfunction, for instance, in a very global nature there is aging, and aging does lead to some dysfunction and it can go hand in hand with autoimmune disease. So if you look at them in a combined fashion, it could lower immunity. However, if I'm looking at things, now, of course my experience is not in immune deficiencies, so that's, it's something that's not really in the remit of what I normally do, so I can't answer that excellent question with a great certainty. However, it's not impossible. That's what I would say. We do see phenomena [that] have an impact on immunity, and it can cause deficiencies and or dysfunction. But I don't really have any studies to point to that would say that either would GBS or CIDP that there's an immune disorder that's closely tied to it.

00:29:22 Beadon

I would add to that, that there's no evidence that people with CIDP or GBS, have, or are more susceptible to infections or are at higher risk of, a low immune system, and

the caveat to that being the medications that we use to treat CIDP can certainly make them more susceptible to infection by blocking their immune system. And specifically, it's those who are being treated with corticosteroids that we see that in, and then to a, to a degree, those who are on azathioprine or mycophenolate or other agents like that, but the disease state itself of, GBS, CIDP or MMN does not put you at risk of being more susceptible to infection. That's been shown quite clearly.

00:30:07 Grant

Thank you. I think that's a question that'll put a lot of people sort of at ease when they think about their own condition and their susceptibility and things like that, but it's true, it is a question that patients often have and it's good to hear that answer. I have another question for Dr. Beadon. So should CIDP and MMN patients get referrals for physical and occupational therapy just like GBS patients?

00:30:37 Beadon

Absolutely. I think that this is a fantastic thing for their function, for their quality of life and, and to maintain the strength and build strength back. And so depending on where you're living and what services are available, many people won't be able to be followed forever by an occupational therapist or, or physical therapist through the public health care system, but anytime there's a flare, or even if there hasn't been a flare, there's kind of just natural aging that changes some of our function. I always encourage my patients to get what we call a tune-up. So our clinic is lucky enough to have a physical therapist working, a physiotherapist, working with us, and they'll do you know anybody who's newly diagnosed, they'll, they'll spend a lot of time with and then, for our chronic patients, they'll do a check-in every once in a while and update their, their exercise program so that they're doing exercises at home on their own on a regular basis. Things like optimizing balance, optimizing, you know, your walking, occupational therapists can be really, really helpful in adaptation strategies for, especially for things like MMN, where hand functioning can be so severely affected, to have tools to overcome some of the limitations of the weakness in, in hands and other strategies. I think that they're invaluable members of the Allied health team and, and I really do encourage everybody to have an assessment at least, and then hopefully you can take from that assessment some exercises or some strategies to do on your own at home.

00:32:19 Grant

And do you think that it's ever too late to start physiotherapy or occupational therapy?

00:32:26 Beadon

I don't think it's ever too late to start, but the earlier the better for sure.

But there's always something that you can benefit from, even those who have had, you know, even my patients who've had CIDP for 30 years and they've adopted super well and they're very stable. That tune-up can be a real benefit.

00:32:31 Grant

That's good.

The next question is for Dr. Vasjar.

My child has CIDP. What are some ways that parents can talk to their children about pain levels?

00:33:05 Vasjar

Well, thank you for your question and I could talk for hours about the pain assessment in our patients. And I think my colleagues here on the panel, they could discuss the same thing, but in children, something hurts, but the difficult part is to quantify the degree of the pain and in teenagers and you know, maybe from the age of 10-12, we can use the different pain scales that we have, say, the happy face, the sad face, you know, or the scale of one to 10, but it's much more difficult to quantify pain in the preschool children.

So my approach, or our colleagues' approach is to make it relatively simple and go with the, only as opposed to 10, 10 scale measurement, only mild, moderate, severe. When we talk to children, the mild will be something like itch, when you scratch your skin and it's itchy, so it would be a level of a mild pain. The moderate pain would be like a mosquito bite or even a wasp sting and the severe pain would be really, the severe dental pain. If the children that has ever been to [the] dentist, they know what the severe pain is. So this is the way you can probably approximate the level of the pain the child has, we know it's very subjective, so people would, not even children, the teenagers , you know, they may say my pain is 10 out of 10 and they are sitting next to me not crying, not screaming, just you know, chatting. So keep in mind it's a very subjective assessment and even though you think that you know what the level of the pain is, it may not be exactly accurate so, but for the preschool children, I will go mild moderate severe.

00:35:40 Grant

And you mentioned that pain is subjective. Earlier we had a question about gabapentin and other medications for pain. So this is maybe a question for the entire panel. Are the things that you recommend to your patients other than, let's say, medications to treat their pain?

00:36:02 Vasjar

Well, we start with the medication, but we implement behavioural therapy like, but it's a bit more complex. You know, getting access from the pain clinic. And I suspect the same thing will be for my colleagues on the panel, but sure there are ways to go, with behavioural therapy, to help with the pain. I think if anybody else wants to comment.

00:36:34 Beadon

I totally agree.I think we have the pain clinics that we have out here in BC do a lot of work on, you know they do pain classes and talking about cognitive- behavioural therapy for pain, mindfulness and meditation for pain, as well as gentle exercise and other approaches like this and the, the trick, as you already alluded to, is getting access to those places since we've got, you know, very long wait lists for the publicly funded pain clinics, we do have some sort of public-private partnership clinics where you can get some of the, some of the services more quickly but have to pay for them. And so that's a real challenge. But certainly there's, there's lots of great evidence for cognitive behavioural therapy and pain.

00:37:19 Vasjar

And it really helps. So if anybody can access it please go for it, for sure.

00:37:26 Beadon

You're, there are apps that are designed for this as well, which are a little bit easier to access than the formal programs. So that may be something to look into if you're not able to access a formal program.

00:37:41 Grant

And obviously apps, you don't need a referral for, but our patients, do they require referrals for these programs, do they require referrals for coverage? What is usually your approach to recommending those?

00:37:54 Vasjar

Yes, they have to be referred by somebody, specialists for the pain clinic, for example, and it's a hospital-based service in our institution. So it's covered by the OHIP at the hospital there.

I don’t know in BC, but probably the same.

00:38:13 Beadon

Yep, same in BC. I would mention though, that cognitive behavioural therapy can be performed by many different practitioners, doesn't have to be hospital, hospital based and those ones are usually private paid. But anyone who has extended health benefits can usually get some coverage through that. But it's a lot trickier than the pain clinics which have the, all of these services wrapped together.

00:38:42 Grant

Dr. Beaudin, Dr. Picher Martel, is it about the same in Quebec?

00:38:45 Beaudin

In terms of access to pain clinics, it's very difficult in Quebec as well. Obviously, I think it's a bit of a challenge everywhere. It's something that the patients draw extreme benefit from, this multimodal approach has been proven to be very effective. For example, mindfulness therapy, cognitive behavioural therapy, those things are really useful. However, access can be extremely difficult and delayed.

00:39:19 4 Grant

Thank you.

Now I have a question for Dr. Picher Martel. Have we seen adverse events in patients that received a vaccine against COVID-19?

00:39:34 Picher Martel

That is a good question. It's still a hot topic. So if I understand the question properly, you're asking, what's the risk of developing GBS if you take the COVID vaccine?

In the past we have associated GBS with some vaccinations, including the flu vaccine. Now if you go back in those studies, it was an, it's a very, very small risk. One to two people in a million that received the vaccine for COVID. There was some evidence that some of the vaccines, especially the conventional vaccines, not the later developed vaccines, but the Johnson and Johnson that I think, we may have, we didn't have in Quebec and Canada, but so the conventional vaccines were a bit higher risk. But we're only talking about a few more cases per million people that received the vaccine, so the current vaccines that we're using, and we have many, many million people, up to 20 million people that have been, that have been vaccinated and none of them have an increased risk of developing GBS when getting the vaccine. So the risk of the infection, we must be reminded, that the risk of the infection, whether it's the flu or COVID is far higher than developing GBS through

vaccination and I'm not sure where the question was going, but somebody who's already had GBS, the risk of of having symptoms after vaccination, that's something that we have less evidence for. However, we say that, after a few months, three months to a year, depending on the sources.

You shouldn't limit yourselves from getting a vaccine, especially if the initial GBS was not associated to a vaccine. It's a bit more complicated if you think that perhaps the GBS was associated to a vaccine, but after the acute phase, there shouldn't be any contraindications.

00:42:12 Grant

Great. Thank you. And the next question was for Dr. Beaudin. It's a bit in the same vein, but a bit broader.

What could make symptoms worse for somebody who's come out of GBS? Are there any things that could, that could lead to a flare-up?

00:42:32 Beaudin

Well, I think we have to draw a distinction between the normal evolution of the disease. For instance, after a few months, a patient might have some fluctuation in their health status.So we might think, is that as the CIDP in the initial phases, or sometimes it's that years later somebody had GBS and they are fully stable and all of a sudden there's a flare-up. So at that point many things could explain it, there are many things we have to look at. For instance, there is a possibility, of, of over the lifetime, in 5 to 8% of GBS patients will have a subsequent GBS episode, however generally it's an exception.

Because of a sensitivity, there could be underlying symptoms, for instance, tingling, numbness that was still there, but a very low level. Those symptoms can fluctuate, based on other factors. For instance, it could be the case with any systemic disease. People experience disease in a fluctuating manner over the course of their lifetime. You also have to think that there could be a new process as well, for instance, a patient who had GBS in the past that then develops diabetes, or has chemotherapy to treat cancer, or takes another, a neurotoxic medication. They may have a new, an additional neuropathy, now depending on how they recovered from the initial disease, of course, the nervous system was affected, it they may have, you know, recovered fully, but over time we may see that the degradation was sufficient enough for something to flare up in the future, but overall it's quite broad and I can't give you an exhaustive list of all the potential causes over all the patients I see for GBS. When I see them during their last follow-up. I tell them if you have future objectives, or if you have new symptoms or a flare-up of your current symptoms or previous

symptoms then call me back. If the symptoms are significant enough then they should see their neurologist again for reassessment. Thank you.

00:45:22 Grant

Now I have a question for Dr. Beadon.

Can CIDP affect cranial nerves affecting, for example, double vision, cough triggered by swallowing, neck cramping or hearing loss? If yes, is this a rare variant?

00:45:40 Beadon

That's an excellent question. So the first thing I would say is, yes, there are case reports of all of those. Well, actually, I would have to go searching for the hearing loss, but I certainly, there are case reports of all of those other symptoms occurring in the context of CIDP.

But the presence of any of those symptoms would also make me think really hard about whether I had the right diagnosis. So this is, a the, in the, if I see any of those things present, I'd really go back to the drawing board, start again with the history, kind of try and make my way through the whole kind of course of disease to convince myself that it is truly an inflammatory neuropathy or CIDP specifically. The presence of you know, for instance, visual changes or swallowing difficulties, these things can happen in regulatory neuropathies, for instance, they can occur in the context of other inflammatory disorders that can cause changes in the nerves, so things like sarcoid for instance, and I would really be looking for any other atypical features to make me kind of doubt the diagnosis, if you will. So our, our current 2021 CDIP guidelines, that's in, that, that's sort of elucidated in the guidelines as well. So if cranial nerve involvement is there, you really have to convince yourself that they meet criteria in other ways in order to secure the diagnosis.

00:47:09 Grant

Thank you.

And I think this may be our last question. So for Dr. Vasjar, what are the chances of CIDP remission in children?

00:47:22 Vasjar

Thank you for the question. It's the CIDP in children and all the other autoimmune neurological diseases in children, are, relatively, compared to adults, are relatively mild to moderate course, and many of the children being at CIDP in myasthenia or even pediatric multiple sclerosis do quite well and I would say definitely around

30-40% of children with, including CIDP will get off the medication and will be in full, full remission. I don’t follow them after the age of 18, so I cannot say if anybody relapsed after the age of 18, but the chance of relapse will, is obviously higher in somebody who had autoimmune disease in the pediatric age.

OK, but I can't tell you the numbers, because maybe my colleagues will know how many of their patients presented in pediatric age, and then they’re off medication, in remission and relapse later on. It happens. But I don't think, it's maybe 5-6 percent max.

00:48:46 Beadon

I think I only have three patients in my practice who had Pediatric CIDP and I have not seen relapses in any of them into their 40s and 50s, as adults. So it's a very small number. But I agree with what you're saying for sure.

It's really hard to relapse after being in remission. The longer the remission, the less likely the relapse is going to happen.

Thank you.

00:49:15 Grant

Thank you. Thank you. So we maybe have time for one last question for Dr. Picher Martel.

This is a question on MMN. I read that MMN is more frequently diagnosed in men than women. As a woman with MMN I'm wondering why that would be the case. Do you have theories to explain why you diagnose it more in men?

00:49:39 Picher Martel

Well, this won't be a very long answer. Yes, we know there are a few more men, but I don't know that anybody can really give you a clear-cut answer. I looked because I wasn't sure. If there was a theory, I didn't find one, but the figures, the frequency figures are quite old. If you look at the studies, they repeat the same thing 2.7 to 2.3 more men than women, 2.3 times more men than women, so they weren't epidemiological studies. They were cohort studies, and they are known to have biases. So it's unclear.

00:50:27 Grant

Thank you. Very much. Thanks for the answer.

And so that will put an end to our Q&A with our esteemed panelists.

Thank you to all four of you for participating in our Q&A, we really appreciate it. Our patients often come to us with questions and concerns and we all always send them back to you guys. So it's good to have you here to answer those questions live. And I know one of the challenges with our rare diseases is that we often don't have all our questions answered, but we hope that you know we'll continue answering these questions and who knows, maybe one day we'll have an answer for why men are more likely to have MMN than women and all the other questions that we asked. So thanks again, for participating today and thank you for continuing to care for our patients and our families and we really hope that we can continue working with you, for our future events with the foundation.

00:51:33 Vasjar

Thank you for having us.

00:51:38 Beaudin

It was a pleasure. Thank you.

00:51:40 Beadon

Thanks very much for having me. Bonne journée à tous!

00:51:47 Bedford

Hello, I'm Darrell Bedford, the president of the GBS/CIDP Foundation of Canada. This concludes our 2024 virtual conference.

Putting on an event such as this requires a huge team effort. On behalf of the board of the Foundation, thank you to all of our presenters and moderators. Thank you to our sponsors, Takeda, Grifols and CSL bearing. Without their support this event would not be possible.

Thanks to Cencora Innovar Strategies, who partnered with us to provide the market access 101 session.

Thank you to the interpreters who allowed this conference to be accessible in both official languages.

Thank you to the staff of the foundation, not just for everything that they did to organize this conference, but also for their ongoing work throughout the year.

Our conferences allow us to share vital information with patients, their families and their caregivers. You are at the centre of everything the foundation does. We hope you enjoyed this virtual conference and we hope that you are able to join us at a future GBS/CIDP Foundation of Canada event.

Whether it be a Walk and Roll, a support meeting or in person at a future national conference, thank you for joining us and we'll see you again soon.