



My child has just been diagnosed with AFM. What do I need to know?

You can listen to the audio of this podcast at: youtu.be/kDlv3iOjfxw

Rebecca Whitney: [00:00:03] Thank you all for being here today and welcome to the SRNA Ask the Expert Podcast Series, a special edition in collaboration with CDC. Today's podcast is entitled, "My child has just been diagnosed with AFM. What do I need to know?" My name is Rebecca Whitney, Associate Director of Pediatric Programs with Siegel Rare Neuroimmune Association, or SRNA, and I will be moderating this podcast.

[00:00:29] SRNA is a nonprofit focused on support, education, and research of rare neuroimmune disorders. You can learn more about us on our website at wearesrna.org. This podcast is being held in collaboration with Centers for Disease Control and Prevention, Division of Viral Diseases.

[00:00:51] This podcast is also being recorded and will be made available on CDC's and SRNA's websites. During the call, if you have questions, you may send a message through the chat option available with Zoom. We'll do our best to get to your questions, and those that we aren't able to address during our allotted time, we will follow up with you individually after the podcast.

[00:01:12] For today's podcast, we are pleased to be joined by Dr. Sarah Hopkins and Sarah Stoney of Children's Hospital of Philadelphia. Dr. Sarah Hopkins graduated from the University of Arkansas College of Medicine in 2004 and completed residency in Pediatrics and Neurology at Cincinnati Children's Hospital Medical Center in 2009, where she remained on faculty for several years.

[00:01:37] Dr. Hopkins came to the Children's Hospital of Philadelphia in 2014 and became Section Head of MS and Neuroinflammatory Disorders in 2017. Dr. Hopkins has an interest in inflammatory disorders of the spinal cord and directs the multi-disciplinary myelitis clinic at CHOP. She has an interest in acute flaccid myelitis, or AFM, and serves as a neurology consultant for the US Centers for Disease Control and Prevention for activities related to AFM surveillance. She is the site PI for the NIH AFM Natural History Study and the International Pediatric Opsoclonus Myoclonus Ataxia Syndrome Registry. Dr. Hopkins serves as Co-Director of the CHOP/Penn Age Span MS and Neuroinflammatory Disorders Fellowship with Dr. Jennifer Orthmann-Murphy at Penn.

[00:02:29] Sarah Stoney, licensed social worker, graduated from Westchester University with a Master of Social Work in 2014, after she received her bachelor's degree in the same discipline in 2006. She joined the Children's Hospital of Philadelphia's Division of Neurology in 2014. Previously, she worked in the emergency department of Penn Medicine's Chester County Hospital and as a family counselor at the Lincoln Center for Family and Youth. At CHOP, she assists patients and their families navigate the complex medical system, advocates with families and their schools and throughout the community, and provides ongoing support to patients and families in multiple subspecialty clinics.

[00:03:11] Welcome and thank you both for joining us today.



Dr. Sarah Hopkins: [00:03:14] Thank you so much for having us. It's always such a pleasure to be here.

Rebecca Whitney: [00:03:19] Thank you. The goal of today's podcast is to support children and families during a critical and often frightening time after a child has been given the diagnosis of AFM. They've typically already been through so much getting to that diagnosis, and the child and family may be quite fearful of what's next or what this unheard-of diagnosis even means.

[00:03:42] What do they need to know about what is happening, and where do they begin with their questions and finding support? Dr. Hopkins, I thought we could begin with you by asking, what does the family need to know about acute treatments being administered once you suspect or have confirmed that AFM diagnosis?

Dr. Sarah Hopkins: [00:03:59] Well, I think that's an excellent question. Of course, the first one everybody wants to know, you know, what's, what's the thing that, you know, we're going to give, that's going to, that's going to fix the problem. So when I think, the best way to think about treatments for AFM is by understanding what we think, what, what AFM is and what we think causes it, and then to understand kind of where we would want various treatments to act.

[00:04:26] So, so when we think about the spinal cord in general or the brain and spinal cord together, we think about the way the nerve cells work. And you've got the first nerve cell that starts in the brain and goes to the spinal cord.

[00:04:40] And, you know, if you've heard Dr. Greenberg discuss this, you know about the first wire and the second wire. So the nerve cell that starts in the brain then goes to the anterior horn of the spinal cord is that first wire. And then in the spinal cord, that first wire connects with a second wire. That second one is the motor neuron that then goes out to give innervation to the muscles in different parts of your body. So when you have a problem that involves that first nerve, or we would call as neurologists, the first order motor neuron, you have problems that are things like weakness, but also increased muscle tone and increased reflexes in the leg when the neurologist tests it with a hammer.

[00:05:29] Whereas when you have a problem with that second wire, or the motor, the second order motor neuron, then you have low reflexes, you're, you're floppy or flaccid, and also have weakness. And the pattern of weakness can be slightly different. So, when we think about AFM, what we think is happening is that a virus, and we think that at least the epidemics of AFM that we have are associated with enteroviruses, then the virus attacks the interior horn cells, which are the beginning, kind of the, those second wires. So that's why our AFM patients get flaccid or floppy weakness. And we think that the virus actually infects the anterior horns.

[00:06:20] That's what we see in, in mouse models of AFM and what we've seen in older kind of autopsy specimens from people that have had AFM. Fortunately, we don't have those from, from recent outbreaks, but the older information that we have suggests that it's an acute infection of the spinal cord. Now, what confuses things is that when you have an infection of your anterior horn cells, your, when you have an infection like this, your immune system is going to try to take care of the infection.

[00:06:54] And sometimes that inflammation that's generated can cause actually injury to the first wire, which can also give you some, some symptoms of higher reflexes in some extremities. So the more common thing will be something like if you're going to have that, the first wire involved, you know, you might have kind of the second wire involved in your arms. And then the longer, first wire that goes down to your legs, you know, you might be floppy in your arms, but then have increased tone in your legs.



[00:07:31] So we've got the problem of the direct viral infection of the anterior horn. And then we've got potentially the problem of immune system over-activation that might cause kind of additional problems. So, in thinking about the acute viral infection of the cord, ideally we would have an anti-viral that worked directly on an enterovirus.

[00:07:55] That's something that we don't really have. We tried with fluoxetine. If you remember, in 2016, we thought that maybe, we thought that because fluoxetine inhibits the virus in a, in a dish, maybe it would work to give it to, to people when they have AFM, and that didn't really pan out, unfortunately.

[00:08:16] But one way that we try to treat the viral infection is by giving people IVIG, which we know includes antibodies, to circulating in our own virus strains. So, so the idea is you want to give people those antibodies as quickly as you can, to help fight the virus, to help fight the viral infection. And then if there's evidence of kind of bystander inflammation from your immune system. So maybe, maybe the patient also has high re-, high reflexes, or you see some involvement to the white matter rather than the, just, just the gray matter of the spinal cord, then you think about whether you might want to give steroids, which are kind of the, the hammer of the immunology world.

[00:09:03] You know, they really tamp down the immune response in all settings. And then plasma exchange, which of course washes out antibodies that may be causing additional problems with the spinal cord. Although, it's important to keep in mind that also washes out the IVIG, so it's important to just think about all those, all those eventualities.

[00:09:25] So those are the main treatments that are used. The most common one you'll see is that, especially at the onset, people trying to inhibit that viral infection with IVIG. And then based on the situation, steroids or plasma exchange may be considered, but that's very patient-dependent.

Rebecca Whitney: [00:09:45] Okay. And is, so there's obviously an order that would need to be followed, but it is dependent upon what is found is going on with that individual patient.

Dr. Sarah Hopkins: [00:10:01] And also it depends a little bit, what else is in the differential. You know, sometimes we see a patient for the first time, and we're sure that they have AFM. But sometimes we're not sure if they have AFM or if they have transverse myelitis or if you've heard of MOG-associated demyelination, the treatments for those things differ.

[00:10:19] So it's important to have a neurologist who can talk to you about all those different eventualities, and why, kind of, they're recommending what they're recommending at the time.

Rebecca Whitney: [00:10:30] To make that differential diagnosis, because the acute treatments may differ.

Dr. Sarah Hopkins: [00:10:36] Yep, depending on the clinical situation.

Rebecca Whitney: [00:10:40] Okay. Is there generally a preference of one over another? Anything that... do they have different goals?

[00:10:51] You touched on that a little bit about, you know, like IV steroids, tampering down that immune system. Among the three that you discussed, do you have a preference one over another that typically works better, or you see better outcomes with it?



Dr. Sarah Hopkins: [00:11:07] Well, so the, so another thing that actually I probably should have mentioned with the first question is that we don't have any treatments that are proven through a study to work for AFM.

[00:11:20] And that's for a lot of different reasons that we may get into, may get into later. But so there's not kind of a prescribed, definitive, 'this is what you must do in AFM,' or 'this is the, the order in which they must, must be given.' So again, it's really just dependent on the clinician's interpretation of the, the presentation. But there, I would emphasize that, you know, there should always be a reason and they should always be able to discuss that reasoning kind of with, with the patient's family.

Rebecca Whitney: [00:11:53] Okay. Excellent. That's very important for families to be able to ask and understand why one may be given over another and what the goals of those acute treatments are.

[00:12:06] How do you know, once you decide what that acute treatment plan is going to be, how do you know if it's working? What should a family expect to see? I know sometimes it can be difficult because it can take time to see results. How long does that wait go on before you may try another one of the available acute treatments?

Dr. Sarah Hopkins: [00:12:33] Well, and that's, that's a fabulous question and I wish I knew the definitive answer. You know, I think that when, at least when I think about giving IVIG, I'm thinking about trying to do what I can to help the immune system keep the infection from getting worse as kind of the first and most important step.

[00:12:59] There is not, as we've already talked about a little bit, a lot of evidence that any of these treatments make people significantly better, especially immediately. They, they all do take some time. And when I think of IVIG, I'm thinking about, you know, can I stop, can I stop things from getting worse?

[00:13:21] You know, these patients are, our patients with AFM will often get worse over the first, you know, kind of five days or so. And I think if we can shorten that period, that that is really, really helpful. And then, sometimes if there is secondary inflammation and involvement of the white matter tract, which is kind of that, which is sensory tracts as well as kind of that, that first wire. You know, sometimes then we will see, we will give later, kind of after the, the acute infection period, steroids or plasma exchange. And that additional inflammation, when that additional inflammation, it goes down, that can, that can get you some more clear quick, quick improvement.

[00:14:15] But again, not everybody has that second nerve, kind of, involvement. So, so it's hard to say, you know, if you don't see improvement after, you know, three to five days, then, then you would need to do another treatment. It's just not as clear cut with AFM as it is for some of our other neuroimmune disorders.

Rebecca Whitney: [00:14:34] Okay. And are some of these treatments, can they be repeated? Do they need to be repeated? So if you gave IVIG, and you went through one course of it, is it possible to repeat it again before trying something else? Is that ever something that comes up?

Dr. Sarah Hopkins: [00:14:55] So, so typically, AFM is something that is kind of a one-time thing. Actually I'd say typically, always. I don't know of any, anybody that's ever had two episodes of AFM. That would suggest to me that maybe you'd want to look for, look for a different cause to the second episode in particular. But for if, for the IVIG, I mean, if you've been trying to intervene with an infection, then, you know, it makes sense that that would be helpful during the acute infection and not so much something that you would see an improvement with later.



[00:15:32] So, so we typically will only give IVIG once, there, with, you know, potentially some caveats depending on, on presentation. But almost always it's only one dose of IVIG.

Rebecca Whitney: [00:15:45] Okay. And that, and just to clarify too, that was my, my question, was in the acute setting, if, if it would be repeated before moving on to, to additional.

Dr. Sarah Hopkins: [00:15:58] So not usually, with the caveat that dosing regimens vary. So the, the dose is usually two grams per kilogram. At some sites, we'll divide that up over two days or sometimes even four days. But that, we consider that two grams per kilo, no matter how many days it's given over, to be one dose.

Rebecca Whitney: [00:16:20] Okay. All right. And what if a family or child isn't experiencing or seeing improvements over the course of these acute treatments? How, how do you approach that? Like, what is the next step? What, what questions should they be asking?

Dr. Sarah Hopkins: [00:16:42] Well, so really, I think when, when a child is first diagnosed with AFM, of course, you know, as your, as a parent, your mind goes to, you know, okay, so how are we going to fix this?

[00:16:56] I, I totally understand that. But because with AFM, we don't have a treatment that is specifically proven to make things better, we have found over, over the past few years since we first started seeing this in 2014, that really the rehab and the physical therapy, the occupational, the speech therapy is really, honestly, probably more important than, than the acute treatments that people are getting when they first come into the hospital. So starting the physical and occupational therapy as soon as possible when those patients are in the hospital, really thinking about what are we, what are we going to do rehab-wise to help this patient get better?

[00:17:45] Those are really key things, because patients can even have improvement, you know, months and months out to their functioning with, with increasing rehab therapy. And then there are also some surgical procedures that we think about usually around six months after onset. Things like nerve transfer surgery, and then a little bit later, usually like a tendon transfer surgery to help improve those, that function.

[00:18:11] So there are other options and it's important to be thinking about getting rehab and therapies really early on in the course, especially with AFM.

Rebecca Whitney: [00:18:21] Okay. Excellent. Thank you. And switching gears a little bit to determining what is happening, coming to that diagnosis. Obviously, an MRI is something that nearly every child with, with AFM goes through. How often are those MRIs done during that acute setting?

[00:18:43] Do you give the acute treatments and then do another MRI to see what's happening in the spinal cord? What should families expect as far as imaging is concerned?

Dr. Sarah Hopkins: [00:18:55] Well, so I think that is one thing that varies a little bit between centers. It's important to realize that even when you're really suspicious of AFM, you know, sometimes an MRI of the spinal cord that is done very quickly after the onset of symptoms can look normal or close to normal.

[00:19:14] So it's not unusual that if that, that first MRI is normal, we will repeat that in five to seven days. And then, for, at our center at least, how frequently somebody is going to get scanned will depend on how they're doing. You know, we really do try to treat the patient rather than treating the imaging.



[00:19:37] So if, so if the patient is getting better, there might not be a repeat image or, or if the patient's stable. Now, if the patient is getting worse, you know, typically there would be repeat imaging probably at about a week or so. So I think, but I think that would vary a little bit by center and based on the clinical presentation of the patient.

Rebecca Whitney: [00:20:00] Okay. All right. And any other interventions or measures that may need to be taken to support a child while they're going through these acute treatments that are important for families to, to consider or to be aware of?

Dr. Sarah Hopkins: [00:20:17] Absolutely. And I think, I think really that supportive care is really, really important for families and providers to be aware of.

[00:20:26] So the first thing is that AFM, especially AFM that involves your arms or that is associated with trouble, trouble swallowing, or weakness of any of the muscles, any of the cranial nerves. So the muscles that go... The nerves that supply the muscles that go to your face. Those patients that present like that are at higher risk for having lesions in their upper spinal cord.

[00:20:52] So their cervical spine. And that means they're at higher risk to have trouble with breathing. So really important for families and providers to be aware that they need to be prepared to monitor breathing with pulmonary function testing, and also that they really need to be prepared to, to intervene if a child is having trouble breathing, because that really is the thing that makes the difference in the child and the child's safety.

[00:21:20] Other things to think about are bowel and bladder issues. Typically, our patients with AFM don't have long-term bowel and bladder issues, but they may have transient issues early on. So things like bladder scans and making sure we're appropriately managing constipation, which is common, you know, when your muscles, when your muscles of your trunk are weak. You want to really make sure that you're helping things move through.

[00:21:47] Other things, sometimes if they're breathing trouble, we'll do things like ultrasounds of the diaphragm to make sure, or to see if we can figure out, you know, is one side of the diaphragm weak, is there anything, is there anything we can do to support that? So, so that's the kind of supportive care to really, to really be looking for, to make sure your child is, is getting.

Rebecca Whitney: [00:22:10] Okay. And if, if a family is not currently seeing those additional care team members like a pulmonologist or urologist, should they be asking if, about it? Or is that more of the, the nursing care team that is going to be monitoring and bringing those folks in?

Dr. Sarah Hopkins: [00:22:34] So, so it depends a little bit. Those are people that you should be seeing for sure if there's any kind of a problem.

[00:22:43] But if, for instance, you're weak just in the leg, it may be that all your pulmonary function testing is going to be normal and you'll never, you'll never have need for a pulmonologist. But it, it is super important that they, you know, keep an eye, keep an eye on all those things so that they're aware if they need to consult those physicians.

[00:23:06] In some centers, they will always consult an infectious disease specialist as, as well. Because that's the person that will facilitate kind of extra testing. If samples need to be sent to CDC. If, you know, if there are special tests for infections that need to be done, they'll help facilitate that.



Rebecca Whitney: [00:23:27] Okay. And that kind of leads me into my next question about specific tests that a child that, who is suspected with AFM or diagnosed with AFM can expect to go through, whether there're imaging studies or, or blood tests to help confirm that diagnosis, to get to the bottom of what's going on.

Dr. Sarah Hopkins: [00:23:49] Well, so there are tests that help confirm the diagnosis, and then there are tests that help us figure out why the person has that diagnosis.

[00:23:58] So one of the most important ones, when we're talking about infection-associated AFM, like the, the bumps in cases of AFM that we, well, we were having every couple of years until social distancing with coronavirus in 2020. So those bumps, it's, it's really important to try to identify if there's a virus associated with it.

[00:24:24] The problem is that you're most likely to find the virus associated with it the faster that you do the testing. So what we actually have set up to do here with the protocol that we have in place moving forward is that when a child comes to the emergency room and we're concerned that they may have AFM, they'll just get the viral testing then. So that viral testing is a nasopharyngeal swab. If you've had the COVID testing that feels like it's, you know, going to touch your brain, it's the same kind of swab. So that is, that is miserable.

[00:25:01] And then, it's also looking for enterovirus in your, in your blood, and we'll also do stool testing to look for enterovirus. And also we'll typically do a spinal tap because we're looking for, we're looking for cells in increased protein that would be consistent with a diagnosis of AFM.

[00:25:24] So, you know, like we talked about, sometimes we do that MRI of the spinal cord at the beginning, and it is normal or fairly mildly involved. So we're looking for kind of additional supportive evidence with that, with that spinal tap. Another test that we may consider if there's a question based on that, on the MRI and the spinal tap is then a test called an EMG where we give little electrical impulses to test, test the integrity of the nerves.

[00:25:56] And sometimes there are kind of needles, needles in the muscles to look at the muscle response to kind of nerve stimulation as well. That is a test that is typically done to kind of verify that it's your anterior, anterior horn, so that, so that the beginning of that second nerve that's involved. It's kind of done to confirm that involvement.

[00:26:23] So that can help clarify the picture too.

Rebecca Whitney: [00:26:26] Okay, thank you. Thank you very much. And kind of switching gears a little bit to, you know, having a child in the ICU is obviously very traumatic and, and, and kind of bewildering experience, having been there myself. And it can be very logistically and emotionally challenging.

[00:26:49] So, Sarah Stoney, how, how does social work come into the picture and help support a family and child during this acute phase?

Sarah Stoney: [00:26:59] So I, I always joke with my patients and families and say that you don't really know what a social worker can provide until you ask them. So never hold back. Like if there is a challenge that you're having, one of the first people I feel like when you're admitted you should try to speak to is a social worker, because they often have access to all the different resources that are going to be available to you through this acute admission. So for example, at CHOP, we have services in our intensive care unit and throughout the hospital that can really help support not just our patients, but also the entire family, including sometimes, you know, clinical psychologists.



[00:27:39] We have child life specialists that can work with our patients and our siblings of patients, and chaplaincy, and of course, social workers as well. One of the things that social workers can help with, and I'm glad you mentioned it, is that aside from this extremely traumatic event of having a child hospitalized, life still continues to go on.

[00:28:00] So log-, logistical things still need to be taken care of, bills need to get paid, people need to be excused from work or school or whatever the case is. I feel like that's one of the strongest areas where social workers can really swoop in to try to take some of that off of the parents' plate and be able to help facilitate whatever that may be, whether it's helping with FMLA paperwork, we're assisting with financial assistance.

[00:28:26] Every hospital's a little different, but there are certainly supports there and are available and likely best to go through a unit social worker for that.

Rebecca Whitney: [00:28:35] Okay, thank you. So definitely something to keep in mind that you're available for, for the full family, including those, those logistics. So, and are there community and support resources in particular that a family should expect that a social worker may help them with locating, whether it's additional insurance or any other resources that you can, can think of?

Sarah Stoney: [00:29:05] A hundred percent. Actually this list would be quite long. And it, a lot of it varies by where our patients are located and what services and organizations are available where they live. Regardless of where you live, the number one thing that you would want to do after receiving this diagnosis is inquire with your hospital social worker about whether or not your child is eligible for Medicaid or medical assistance.

[00:29:32] So every state, unfortunately it's not a federal program, but every state has a different medical Medicaid program for children under the age of 18. And in many states, just the diagnosis or level of disability can qualify a child for medical assistance. And why that's so important, particularly for families that are at the beginning of this AFM journey, is really because that, that secondary insurance will scoop up everything that the primary doesn't cover.

[00:30:03] And, and help cover copays deductibles and saving the family thousands of dollars in the long run. It's not available in every state, however, usually in the states where it's not available, there's waiver programs or there's other available resources. So even if there isn't a specific Medicaid program in your state, there's likely some sort of financial offset to your medical expenses that they would be able to help you find.

Rebecca Whitney: [00:30:28] Okay. And that, that leads me to, even if a family has private insurance, it's important to still inquire about these possibilities because they could still be eligible based on the diagnosis or, or potential disability. And the social worker is someone that can complete that application or assist them in completing that application.

Sarah Stoney: [00:30:52] They can absolutely assist them with that application. One of the key things that, that we do at Children's Hospital is that we prepare all of our families with a letter from our clinic very clearly outlining why this child is eligible for medical assistance. That's very helpful. So if you are working with your team, with your medical team on this, ask them for a letter. It makes going through the process a lot simpler.

[00:31:21] And I have, you know, this conversation with families all the time. Applying for any type of assistance, particularly through a government organization, just like doing your taxes, it's not fun by any means. And it's very, very taxing and challenging. But in the long run, it is, it will be so beneficial from a financial standpoint.



[00:31:42] We talked earlier, Dr. Hopkins mentioned about the need for, the real critical need for ongoing rehab, such as PT, OT, and other therapies, and getting that Medicaid or medical assistance could be your ticket to, in order to not allow those visits to get capped yearly. So with, we do have some patients that unfortunately are not eligible for secondary insurance or Medicaid, and we typically try to advocate and fight for additional visits based off of the diagnosis.

[00:32:14] Thankfully there is literature out there that supports that, but if you do have the secondary Medicaid, those visits would be unlimited.

Rebecca Whitney: [00:32:23] Okay, thank you. And talking about the, the rehabilitation services that, that hopefully are going to begin while they're still in that acute phase, because it's so critical to their recovery.

[00:32:41] Are you also able to assist, would it be you that, that a family would go to, to help them kind of coordinate what happens beyond that acute phase? Once they move off of that ICU onto a regular med surge floor or to an inpatient rehabilitation hospital that may be at a different site. Can you talk a little bit about that process?

Sarah Stoney: [00:33:07] Yeah. So one of the things that I think is so interesting about CHOP is, is kind of everything that goes on behind the curtains that maybe families and, and patients don't necessarily see. But the amount of coordination and communication that happens on nearly every patient in the hospital is just absolutely extraordinary.

[00:33:27] So it's a combination of, you know, not just myself, but the medical team both inpatient and outpatient. So while Dr. Hopkins may be consulted to see a patient with a potentially new diagnosis of AFM, she is an outpatient physician and may only be consulted to due to her level of expertise.

[00:33:50] So outpatient, inpatient teams, psychosocial teams, nursing teams, everyone kind of collaborates together to decide on what the plan looks like. Where the social worker's role comes in that we do assist with the execution. So once we know what the plan is - is it locating, you know, rehab facilities that are within driving distance for the family or helping to make those appointments?

[00:34:16] Those are certainly things that we can assist with.

Rebecca Whitney: [00:34:18] Okay, thank you. And something else that comes up often in the acute setting is families may be being asked to participate in research. They're already being inundated with, with information, new information, and it may seem like a peculiar time to be approached about an okay to participate in research.

[00:34:44] Dr. Hopkins, can you explain a little bit about why they may be being asked to do so during such a critical time and maybe a little bit about the role that research has played and continues to play in AFM?

Dr. Sarah Hopkins: [00:34:58] Absolutely. I'd be happy to. So like we talked about a little already, there are lots of unanswered questions with AFM.

[00:35:07] There are questions about what's the best acute treatment. What are the, when should you be getting the treatments? What are the therapies that make the most difference in outcome? There are also kind of some, some potential new treatments in the pipeline, like antibodies that would be specific to an enterovirus called enterovirus D68 that we think is one of the main ways, one of the main causes of AFM.



[00:35:31] So there are a lot of, a lot of ongoing questions to be answered. And the hardest thing with AFM has really been that the cases occur kind of every couple of years and they occur in different parts of the country. So, so it's really been difficult to study kind of for that reason because no one center has a ton of cases, and also, different cases present differently.

[00:36:03] So one, one patient may present with arm weakness. One may have bilateral leg weakness, might, one might be weak in all four extremities. And we don't know what the predictors are for how they're going to do. So, there is now the NIH AFM natural history study, which has centers throughout the country.

[00:36:23] So starting now in enrolling new patients in those studies will help us, help us to answer all those questions and really is a massive contribution that families, families can make to, you know, upcoming patients that will have AFM. But also they're even helpful in many cases for, for your own child, you know. Like we talked about, there's, there's often some question, you know, are we a hundred percent sure that this is acute flaccid myelitis?

[00:36:54] Or could it be transverse myelitis or MOG-associated disorders? So, so for instance, when, when a patient is enrolled in the AFM natural history study, there is an adjudication committee for patients that come in. You know, there's a committee that will review MRI scans and things like that. So, so that actually is a good resource for your physician to help, you know, ensure that, you know, we're considering all the different ins and outs and we've got, we, we're sure we've got the right diagnosis and that kind of thing. So it really is about not only identifying the cases and capturing them so we can learn more about it, which has been hard up until now, but also about kind of maximizing care for each kind of individual child.

Rebecca Whitney: [00:37:45] Okay. Thank you. And I know you talked earlier about some of the tests as well and samples, and I know that that is also a big piece of, of the research is, is collecting those different samples so we can help identify.

[00:38:01] And CDC has, has their studies and different centers including, including organizations like SRNA have, have studies as well. So, not participation is, is critical to help us for future generations.

Dr. Sarah Hopkins: [00:38:21] Absolutely, absolutely.

Rebecca Whitney: [00:38:22] Yes. And another question. Sometimes we have caregivers who are nervous to ask their physicians questions. They may be in a position where they feel like they're not the experts and they're not okay to do so. But I think it's, it's critical for families to be able to ask those questions and to have a good rapport with their physician, because that is their child.

[00:38:49] That is their baby that is going through this. Do you have any advice for families, both from the physician perspective and, and Sarah Stoney from, you know, from the other side of it as a social worker supporting the families, as far as how to communicate with their care team, how to go about asking the questions, or what to do if they are experiencing a difficult time with those communications?

Dr. Sarah Hopkins: [00:39:18] So, so I think that the first thing is that I feel like, I feel like I'm always telling families that, you know, you're the expert on your child. And I think that is, that is absolutely true. You know, I've seen, I've seen more AFM than you have, but I don't, I don't know your child. So, so I need you to, we need, we need absolutely families to work with us so that we know kind of all the ins and outs about what's best to kind of do for their child.



[00:39:48] And, and I would, I would hope that, you know, there, there would be a dialogue, a dialogue, both ways that, you know, the physician is more than happy to answer questions if they know that, if they know that they're there. So I would really encourage you to just keep asking if you're not getting an answer, but also to pull in other members of the team.

[00:40:17] So for instance, if you're, if you're not getting an answer from one physician, then, you know, reach out, reach out to the nurse, discuss your concerns there, reach out to the social worker. But we, we really try here to make sure that we're sitting down with families on a regular basis and, you know, that's one-on-one, but that's also, you know, sitting down for a family meeting, you know, maybe after the first, after the first week with all the different specialties, specialties, so that we're sure that, that the family is getting their questions answered.

[00:40:51] But then there are also wonderful external resources also like the SRNA and the AFM Family Association. So there are also other resources families can reach out to.

Sarah Stoney: [00:41:06] Yes. I absolutely echo what Dr. Hopkins said in terms of very much so understanding that parents are the expert of their, of their child.

[00:41:16] So while Dr. Hopkins and our teams bring a level of expertise, our, our parents do too. So that collaboration is absolutely critical. So without their critical thinking and their questions we're not going to be able to really fully be able to do our job. So a tip that I often give to families is that we completely understand and recognize that a lot of what we're talking about in clinic appointments is overwhelming, confusing, and often said in language that is hard to understand.

[00:41:50] And we understand that. So we try to keep lines of communication as open as possible. So my suggestion to families would be, you know, when you meet your new outpatient physician or team, figure out who your players are. Who do you call for refills, who do you call if you have medical questions, or just other general support. Know, know who your key players are on your team.

[00:42:15] And then additionally, you, you might be washing the dishes one night and you think of a question of something that you forgot to ask the doctor, keep a note pad around you, maybe in a centralized location near the phone or in the kitchen, someplace where you can jot those questions down. Because even though in that moment, you'll say, 'Oh, I'll remember the ask Dr. Hopkins that on Friday,' you likely won't, and that's okay.

[00:42:40] But we want to make sure that that question gets answered. So try to keep a pad around where you can just jot down those questions and make sure your questions, you get to, an opportunity to ask those questions throughout, throughout your visit. We certainly don't want you leaving with anything additional on your mind. If we could potentially either answer the question for you or be able to, to take the burden of looking into it off your plate.

Dr. Sarah Hopkins: [00:43:08] And we often will ask families to do that as an inpatient as well. You know, questions will come up and then families will say, 'But I'm going to think of more as soon as you walk out the door,' and that is 100% true.

[00:43:22] And we're not going anywhere. We're there all week usually. And when we're not there, somebody else is there. So please, please just keep a running list of questions. And we're happy to sit and go, go through those with you. And, And

Rebecca Whitney: [00:43:40] yeah.



[00:43:41] Yeah, I think it's important to, to remember that there's, there's no question that should be off the table when it comes to, to your child and, and what you're, you're going through, and, and always encouraging.

Dr. Sarah Hopkins: [00:43:56] And if you feel like there is, then that would be a concern for me.

Rebecca Whitney: [00:44:01] Excellent. Good to know. Thank you both so much. And we did have a question, too, come in from our community. And this one is going to be for, for Dr. Hopkins. And it's what work or studies are in the works about developing a treatment protocol for AFM. Is there a suspected link between the effectiveness of PLEX in the earliest detection?

[00:44:28] In other words, would it make sense that PLEX is most effective while the virus is still actively attacking the spinal cord? Because you're giving the body antibodies to attack the virus and steroids work better after the virus is gone, and the immune system's inflammation is causing the secondary damage?

Dr. Sarah Hopkins: [00:44:47] So, again, going back to what we talked about at the beginning, with the first wire, the second wire, where you're looking for treatments to work, the treatment that hypothetically you would think would be most likely to help with an acute infection would be IVIG, where you're giving antibodies that... and we know that IVIG typically does include antibodies to circulating enterovirus types.

[00:45:13] So that would help kind of to treat an acute infection, potentially. At least that's the theory behind why we use it. And then of course, PLEX would actually, plasma exchange would actually do the opposite. It's washing out antibodies. So, hypothetically, there may be, there may be times when, you know, kind of at the beginning, you, based on the clinical situation, you may want to, you may want to give IVIG first and then hold steroids and PLEX if the patient is very sick with kind of an acute infection and consider giving those later if there's, if there's residual.

[00:45:54] But again, very, very based on each individual patient. But in terms of treatment protocols, I really do hope that that's something that some of these ongoing studies are going to be able to help us with, to try to standardize some of the evaluations for patients when, when they first present so we're getting all the same information about those patients, so we can help predict, you know, who needs, who needs what, and, you know, kind of in, in, in what order and how effective is that likely to be. And the other thing is some of this probably has a time impact. You know, we know from, for instance, the mouse model studies that, you know, if the mice get, if the mice get the IVIG really quickly, then they do, then they do better.

[00:46:43] But, you know, one of those studies was 24 hours or less. And sometimes we, we aren't able to give it that quickly to our patients with AFM. So there are a lot of questions still that remain to be answered about treatments, but I'm hoping that some of these bigger studies with some of the standardized data collection will start moving us towards some of those answers.

Rebecca Whitney: [00:47:06] Okay, thank you very much. And it, it reminds me to, to also when, when parents are asking their questions of the physicians, that there simply will be some questions that the answer truly is, 'We don't know, and, and we're, we're still looking and we're still, still trying to get to the bottom of it.' We did have another question come in, and that is how do you involve your older child in understanding an AFM diagnosis and educating themselves about what is going on?

Sarah Stoney: [00:47:44] So in terms of educating siblings, a lot of it has to do with the, the age of the sibling that you're, you're interested in providing the education. At a pediatric hospital, we are fortunate enough, and most do, have a discipline of child life specialist. And they are like master's level licensed therapists that specialize in medical play.



[00:48:09] And they work with all ranges of ages from infants, you know, in our NICU all the way up to young adults. And really what their expertise is, is explaining medical jargon and language and medical disease courses in very age-appropriate dialogue and understanding. So they would absolutely be an excellent resource. If there's not a child life specialist available, I would suggest connecting with a social worker, but then also encourage the older sibling, if old enough, to attend a medical appointment for their sibling and to maybe come prepared with their own list of questions about, that they may have - worries, concerns or questions about the diagnosis, so that they have an opportunity to also ask the doctor questions that are coming strictly from their point of view.

Dr. Sarah Hopkins: [00:49:11] I wasn't sure if the question meant older child, like older sibling or an older child who was just diagnosed with AFM. But I think the child life specialists and social workers are a good resource. The child life specialists are very good at explaining ways in kids could understand. But I do find honestly that, that whole, whole discussion that I stole a little bit from Dr. Greenberg - the first wire, the second wire - is that even for, you know, for, for the patient, for children that sometimes helps them understand, you know, where the problem is. And you know, how, how we're going to work with rehab and therapies and things to get better. Because I think, I think that's really important that the child themselves understand that there really is a reason why we're recommending that they do all this, do all this work. That there's, there's kind of a good reason for it.

[00:50:08] I will say, and this is actually one thing that I meant to mention earlier when we were talking about consultants, you know, it, it would be completely normal for a child, a previously healthy child with a recent diagnosis of AFM, to have some trouble adjusting to that. I mean, who would, who would not. It's a huge challenge. So I think keeping that in mind, and also having even a low threshold to get behavioral medicine and our psychologist involved is, is really important. So just important to keep that in mind, too.

Sarah Stoney: [00:50:42] Absolutely. I was going to add that as well, Dr. Hopkins. So if the question was geared for an older child with AFM, in fact, a lot of the same tools that we talked about could be applied. Our child life specialists don't just work with siblings. They do, in fact, work with the patients themselves, as do the social workers. And then additionally, really encouraging them to become a part of their healthcare process.

[00:51:06] You know, at age-appropriate times, encouraging them to come up with one or two questions for the doctor either that, or just completely independently from, from themselves. So that's always a good idea. I think particularly with our patients with AFM, given that there's such an acute phase and then so much patience required for the really long road of rehab that so many of them have that, that it is completely understandable that our children would have challenges coping.

[00:51:39] I'm a very strong proponent in believing that I think that all teenagers pretty much should probably be in some sort of behavioral health therapy, but certainly when you're dealing with a chronic diagnosis and a recovery time, it's absolutely appropriate to have those, those supports in place. And don't worry about, you know, potentially getting those in place a little prematurely, because then there's a, an established ground so that when issues do arise or if the child who has AFM is starting to really have challenges with lingering disability, and it's affecting them mentally, there's already a basis there of where they can go to for support.

Rebecca Whitney: [00:52:18] Excellent. Excellent. Thank you. And thank you for underscoring the importance of having a baseline and the behavioral medicine supports in place as well.

Dr. Sarah Hopkins: [00:52:32] Can I add one thing? And I think also having a provider that the child is willing to talk to. Like there was a recent patient that we have been, I was having, I was having just a little chat with just the patient by myself. And it came out that not being able to play video games was a huge quality of life



issue. But that's something that, you know, it turns out we're lucky to have an occupational therapist that specializes in making video games accessible. So that's something we're really going to be able to help with.

[00:53:09] So having the child have at least a little rapport with kind of the clinical team is very, very helpful in terms of us being able to help maximize quality of life for them.

Rebecca Whitney: [00:53:21] Okay, thank you. And we did get another question. This one is, we now believe my son experienced a spinal stroke instead of transverse myelitis.

[00:53:33] I have trouble using the correct language when speaking to my son's neurologists and rehabilitation specialists. Can I describe his damage as a lesion or are lesions specific to TM or AFM? His damage is from C3 to T2, if that is helpful.

Dr. Sarah Hopkins: [00:53:53] Yeah. So that's a good question. And I think that terminology is, is often, often a sticking point for a lot of families.

[00:54:04] I think if you describe it as a lesion, people will understand what you're, what you're talking about. I think that's a completely acceptable term to stick with, and, and probably what I would, what I would use. Any, anything else just feels a little bit harder to kind of, to kind of communicate. I mean, really what you're dealing with is an area that didn't get enough blood supply and then an area that has edema kind of beyond that, often.

[00:54:41] But I think, I think just describing it as a lesion, absolutely, everybody will know what you mean. And that is that whole spinal cord stroke versus AFM versus transverse myelitis is sometimes, sometimes a very difficult and confusing, confusing one to sort out. So, so I'm, I'm sorry for that, because I know that it is often, often difficult with terminology and things and it can be a challenge. But I think lesion is fine.

Rebecca Whitney: [00:55:08] All right. Thank you so much. And we're actually coming to the end of our hour, and I just wanted to see if there was anything else that either of you would like to add that you'd, that you always try to make sure that families who are newly in this situation, that you want to make sure that they know and that they understand.

Sarah Stoney: [00:55:34] I would, I would just, lastly, since we didn't touch on it too much, lastly, really encouraged families to utilize their medical team when it comes to planning for schools. I often feel as though families aren't aware that that's actually something that we can and should be involved in, specifically since so many of our patients qualify for 504 plans.

[00:55:55] Those are really, really guided by the medical team. So really ensuring that there's clear communication between the medical team and the school, all, which can all be done through either an education coordinator or the social worker. But really making sure that they're a part of that process to alleviate some of that stress off of you.

Rebecca Whitney: [00:56:15] Thank you. And Dr. Hopkins, anything you'd like to add?

Dr. Sarah Hopkins: [00:56:19] I think, I think just really keeping in mind, you know, what we talked about before about not being, not being afraid to ask questions. You know, I think it's really important. You know, you may get an answer of, 'I don't know.' My patients know that I, you know, if I don't know an answer, they, you know, I'll be very clear with that. But I think important to ask kind of anything that's uncertain to you, anything that's unclear about the plan, anything, anything like that at all.



[00:56:52] I just really want families, you know, this is a confusing time, and I really want families to feel like they have the resources to go to, to get those, those questions answered. Because there shouldn't be kind of things nagging at the back of your head about what, did we consider this? Did we do this? You know, you should feel like all of your concerns have been addressed.

Rebecca Whitney: [00:57:16] Excellent. Thank you. And I hope families, too, understand that they're not alone. There are other families that have gone through this and that there is a community of, of medical specialists, of scientists, of parents and other children that are there to support them.

[00:57:39] So if there are any questions for anyone who's listening to this later on, please feel free to reach out at any time.

Dr. Sarah Hopkins: [00:57:48] Absolutely.

Rebecca Whitney: [00:57:49] Thank you both, once again, for joining me today, and thank you for the assistance and the support that you've provided to our community.

Dr. Sarah Hopkins: [00:57:58] Thank you so much for having me.

Sarah Stoney: [00:57:59] Thank you so much.

Rebecca Whitney: [00:58:00] Thank you, goodbye.