

71| Neuropsych Bite: Pediatric Transverse Myelitis – With Dr. Lana Harder

May 15, 2021



This is an audio transcription of an episode on the Navigating Neuropsychology podcast. Visit www.NavNeuro.com for the show notes or to listen to the audio. It is also available on the following platforms:



Speakers: Lana Harder, Ryan Van Patten, John Bellone



Intro Music 00:00



Ryan Van Patten 00:17

Welcome, everyone, to Navigating Neuropsychology: A voyage into the depths of the brain and behavior, brought to you by INS. I'm Ryan Van Patten...

John Bellone 00:25



..and I'm John Bellone. Just a quick reminder that a new batch of NavNeuro episodes have been approved for CE credits through INS. We want to thank Dr. Ben Hampstead, who has been integral in getting these episodes through. We also want to thank Dr. Mark Norman, the CE committee, and everyone at INS who has put a lot of work into this project. We truly appreciate all of their efforts. If you're interested in CEs visit navneuro.com/INS.

Ryan Van Patten 00:52



Today we have our third Neuropsych Bite with Dr. Lana Harder. Today's topic is transverse myelitis in children. Lana is board certified in clinical neuropsychology and pediatric neuropsychology, and she's an associate professor at UT Southwestern.

John Bellone 01:08



One quick note before we get started. Lana mentions that she had an N of 24 children with transverse myelitis, but she was referring to her 2012 paper. She has since seen many more kids with this condition.

Ryan Van Patten 01:21



Because this is a continuation of our conversation with Lana about pediatric demyelinating conditions, we'll just jump right back into the discussion without any introduction or small talk. So, now we give you Dr. Lana Harder.



Transition Music 01:34

Ryan Van Patten 01:43



So, transverse myelitis is a monophasic disease of spinal cord inflammation, meaning it involves a single autoimmune episode or event. Talk about important clinical differences between monophasic diseases, such as transverse myelitis, and chronic diseases such as MS. And then, if you don't mind, tell us a bit about the pathophysiology of transverse myelitis as it presents in children.

Lana Harder 02:07



Sure. So, when we talk about a monophasic disorder, like transverse myelitis, this refers to a one-time event. So we don't expect a recurrence of this over time, generally speaking. This is compared to a condition like multiple sclerosis that's

recurrent, it's chronic, and we do see relapses over time. Some differences between TM and MS, aside from that medical course I just talked about, is that TM is restricted to the spinal cord. In MS, you can have TM, you can have spinal cord lesions, but there's also brain and optic nerve involvement. Those are some of the ways we differentiate the two of those.



John Bellone 02:50

That's a good way to differentiate. Do we know what regions in the spinal cord tend to be most affected by TM?



Lana Harder 02:56

Yes. So when we look at adolescents and adults, we tend to see thoracic regions involved as opposed to really young children which have more cervical lesions. Also, I think really important to note is a variant of transverse myelitis, which you all may have heard of in the news - it's called acute flaccid myelitis. What you might remember, if you did see it in the news, was this outbreak of polio-like symptoms in children. And this started around 2014 and occurred every two years between July and November. So we saw, kind of, this seasonal pattern. It has been linked to a virus, enterovirus D68. But the reason I'm bringing this up, in response to your question, is that acute flaccid myelitis tends to impact the gray matter, that interior horn of the spinal cord, which then leads to a pattern of weakness, of flaccid paralysis. So we have the two wires responsible for motor control - wire 1, which is the signal descending from the brain to the spinal cord, and wire 2 being from the spinal cord out to the muscle. And it's that second wire that is impacted when we see acute flaccid myelitis. But certainly there can be mixed gray and white matter involvement.



John Bellone 04:19

Okay. And yeah, just for listeners to help them also keep this clear, "myelitis" refers to an inflammation of the spinal cord - so, we're talking just spinal cord here. And then "transverse" means extending across something - so across the spinal cord. That may be one way to remember it.



Lana Harder 04:37

Exactly.



John Bellone 04:38

So, in terms of distinguishing TM from something like ADEM, acute disseminated encephalomyelitis, and neuromyelitis optica, the other myelitises [laughs] - how do we distinguish this one from those?



Lana Harder 04:55

Yes, yes. ADEM may or may not involve the spinal cord. So, by definition, the brain is involved. We may also see optic nerve and spinal cord involvement. So ways that TM and ADEM are similar is they are a one-time event. But the ways that they're different is that there is brain involvement in ADEM, but not obvious brain involvement in TM. And then neuromyelitis optica actually does involve TM. So it's sort of TM plus optic neuritis, which gives us neuromyelitis optica. But, again, no obvious brain lesions for neuromyelitis optica. The other distinguishing feature is NMO, or neuromyelitis optica, is chronic and has relapses.



John Bellone 05:41

I see. So, just to recap: there's "myelitis", the word, in both transverse myelitis and neuromyelitis optica. So we know it has spinal cord involvement.



Lana Harder 05:51

Yes.



John Bellone 05:52

With ADEM, there's both brain and spinal cord involvement. Okay, that helps.



Lana Harder 05:57

Yeah, so, for ADEM, there's brain involvement, which is a primary feature, and then there may or may not be spinal cord involvement as well.



John Bellone 06:06

Yeah. I'm not a pediatric person. [laughs] So this is a good review. I reviewed for the board exam, and that seemed to have left me now.



Lana Harder 06:14

[laughs]



Ryan Van Patten 06:16

You co-authored a 2017 review paper with Alexander Tan, and there's a great table in that paper - Table 1, I think - that sort of breaks it all down in terms of distinguishing these different demyelinating conditions. So we'll include that in our show notes, and listeners can refer to that.



Lana Harder 06:31

Excellent. We created that in our clinics and offices when we were teaching this to people just because of the complexity that you're hearing as I try to describe and distinguish these conditions. I think that's a great resource.



Ryan Van Patten 06:44

Yeah.



John Bellone 06:45

Yeah. Excellent. Well, we'll include it in our show notes. In terms of the prevalence of TM in children, do we have a sense of that?



Lana Harder 06:53

We think it's probably about 300 cases per year in the United States.



John Bellone 06:59

Okay.



Ryan Van Patten 06:59

Yeah. TM is more common in adults than children. Correct?



Lana Harder 07:02

That's right. It's quite rare in kids.



Ryan Van Patten 07:06

So, given the spinal cord inflammation, what are common physical symptoms of TM?



Lana Harder 07:11

Yeah, so, of course, this will depend on the extent of damage or inflammation in the cord, and exactly where in the cord this is located. But, common symptoms would

be numbness, weakness, trouble with balance, difficulty walking, changes in bowel/bladder function. Pain is also a common symptom that we hear about. We published a paper where we surveyed a community of individuals who experienced TM, and, for kids, the caregivers filled this out. But, in pediatrics, what we found were they had more common pain or weakness at onset, whereas adults were describing more sensory change at onset. So those are some of the symptoms.



Ryan Van Patten 07:55

Gotcha. And then what are a few common medical treatments given to these children?



Lana Harder 08:00

Yeah, so it kind of depends on where in the course we are. So at the acute stage, during the inflammation, you hear providers talk about "putting out the fire". And so steroids are often the first-line treatment there. There's also plasmapheresis, or plex for short. That is a process - it's cleaning the blood of the inflammatory proteins. That's also used. The intravenous immunoglobulin, IVIG, is an infusion therapy that is thought to reduce or block the abnormal response in the blood. And so those are some of the first-line treatments. As time goes on, in transverse myelitis, our patients are often left with residual difficulties. So those treatments are about managing those symptoms - like pain, or the spasticity they experience. There are a whole host of symptoms that they have to manage over time.



John Bellone 09:05

You had talked before about how there's different areas of the spinal cord that might be affected - cervical versus thoracic - and where the lesion occurs is going to impact the physical symptomatology, right?



Lana Harder 09:19

Exactly. That's right.



Ryan Van Patten 09:19

In terms of symptoms, TM does not obviously or directly impact the brain. So some people might wonder why neuropsychologists and our, you know, cognitive expertise would be seeing these children. But you've published evidence to the contrary - that it does have cognitive effects. So what can you say about that?

Lana Harder 09:39



I will tell you, this was not a hypothesis that I had. I didn't want to test cognitive problems. The way it came about was we were evaluating our clinic data, and we saw that our patients with MS who, you know, are known to have a chronic brain-based medical problem, were having the same rate of school problems as our patients with transverse myelitis. And that really led us to zoom in on the cognitive data that we had. Our goal when we opened the clinic back in 2009 was to screen everyone that came through with our cognitive screener. So we were fortunate to have this data. At the time we had a cohort of 24, which is, you know, a very large sample when you study rare disorders, but, in general, is a pretty small sample size. We saw average means across the measures, but this wide range across our patients in terms of their functioning. The way that we looked at this was we evaluated frequency at certain levels - so, at or below one standard deviation, or one and a half standard deviations, from the mean. And what we saw looking at or below one standard deviation was a high rate of fine motor speed and dexterity difficulties, followed by about 40% having attention problems, and 30% having memory problems. These were a bit higher than the rates we saw in processing speed and verbal fluency. We were interested to look across other factors, just qualitatively to see if the same patients having cognitive difficulties were also reporting depression or were on different medications that could have side effects. We couldn't find a pattern there, again, in a small sample size. But what we did see was overlap of those who struggled cognitively and reported high fatigue. So that really made us wonder about the role of fatigue and cognitive difficulties in the context of transverse myelitis.

Ryan Van Patten 11:40



I see. That was going to be my next question. What might be going on under the hood, in the brain or in a spinal cord, that would lead to cognitive problems if there isn't direct brain involvement? So, it sounds like one hypothesis is fatigue.

Lana Harder 11:56



Yes, absolutely. That might be my leading hypothesis lately. I think another one would be just the inflammatory cascade that occurs in the central nervous system. Could there be some impact on the brain that we just can't appreciate in an MRI image?

Ryan Van Patten 12:12



Right. And again, that was going to be the next question. Brain MRI studies have not found any differences in TM compared to controls? Is that right?



Lana Harder 12:22

Not at this time, that I'm aware of.



Ryan Van Patten 12:24

Yeah.



John Bellone 12:26

I'm curious how often you see children with TM in your clinical practice and what the neuropsych eval might look like, in terms of referral question, interview, test battery, feedback?



Lana Harder 12:39

So this, in addition to MS, is one of our largest groups in the clinic, so I see them quite often. In contrast to multiple sclerosis, where we're managing what has been described as an "invisible disability", in transverse myelitis, they're often these visible features of the disability. Frequently our patients use a wheelchair or crutches to navigate their environment. And, as I've had patients and families point out to me before, people may make assumptions about their cognition based on these things that are visible. And usually these are the wrong assumptions, right? I know a lot of times our teens and even school-aged children are struggling to manage this and navigate this in the social setting at school. A lot of times I hear them say that people won't talk to them directly, they'll talk to their parent instead. So we do work a lot on self-advocacy and coping. And then also how to manage these residual symptoms that they are managing on a day-to-day basis. So we work on implementing school accommodations to help address difficulties with fatigue and pain. A lot of work is surrounding transition planning, and helping our patients be as independent as possible as they grow and develop and make plans for the future - college or work and so forth. So those are some of the, I would say, "themes" that we address with our transverse myelitis population.



John Bellone 14:17

Yeah, I'm glad you mentioned the social aspect. I can imagine maybe they would be more likely to be bullied at school. I guess our psychosocial skills and our intervention skills probably will be more at work than our assessment skills for most of these kids. Has that been your experience?

Lana Harder 14:35



I would say the differences in their social experiences really stand out to me compared to kids that I work with where the disability may not be as readily observed. And so we talk about what that means and how to manage that. When I participate in the camps for kids with these disorders - I run groups with our teens with transverse myelitis and also our school-aged kids - and it's just incredible to hear them share with each other how they manage this. I think that's a really rich source of support for them to hear from their peers on that. So yes, we're very comprehensive in our evaluations, just like we probably all are in neuropsychology. In addition to that cognitive piece, we're really diving deep into the psychosocial realm and making sure that they have the tools they need to cope and manage, and be as independent as possible.



John Bellone 15:32

Great.



Transition Music 15:33



Ryan Van Patten 15:38

Well, that does it for our conversation with Lana. We have two more upcoming pediatric Bites with her on acute disseminated encephalomyelitis and anti-MOG associated disease. So stay tuned for those. And, as always, thanks for listening and join us next time as we continue to navigate the brain and behavior.



Exit Music 15:58



John Bellone 16:21

The Navigating Neuropsychology podcast and all the linked content is intended for general educational purposes only, and does not constitute the practice of psychology or any other professional healthcare advice and services.



Ryan Van Patten 16:33

No professional relationship is formed between us, John Bellone and Ryan Van Patten, and the listeners of this podcast. The information provided in Navigating

Neuropsychology in the materials linked to the podcasts are used at listeners' own risk. Users should always seek appropriate medical and psychological care from the appropriate licensed healthcare provider.

End of Audio 16:51