Optimizing Occupational Participation and Performance for Patients with Amyotrophic Lateral Sclerosis

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- [Fawn Carson] Today's course is Optimizing Occupational Participation and Performance for Patients with Amyotrophic Lateral Sclerosis. Our presenter today is Scott Rushanan. He is the current director of Rehabilitation Services and co-director of Penn Medicine At Home, part of the University of Pennsylvania's Health System. Scott previously spent eight years as the lead occupational therapist at Penn Medicine's ALS clinic. He is currently pursuing his Post-Professional Doctorate in Occupational Therapy at Colombia University, focusing on cognitive impairments for patients with neuromotor disease. Welcome, Scott, so glad to have you.

- [Scott Rashanan] Good afternoon, everyone. It's wonderful to be here. Okay, so today we're going to be talking about a neurodegenerative disease that I believe we as occupational therapists can play a very impactful role on improving the lives of patients who have it. As we go through the course content, all right, the learning outcomes. So, at the end of the course, you will be able to describe the signs of ALS and the resultant loss of physical and cognitive function associated with the disease, describe the environmental, social, and person factors that may impact function and participation in patients with ALS. And we're going to go over quite a few strategies and interventions that are occupational-based in occupational therapy theory, meant to optimize occupational participation and performance, but by the end of the course you'll be able to at least describe five of them. As we go through the content, I think you'll realize that this is a very diverse disease process, and there's a lot of content to cover here. If you find that there's anything in here that I go over that you want more detail about, I do encourage you to do some self-directed learning and do your own research about it. There's just not enough time in an hour to go through everything in the amount of detail that you may want.

So, I'm going to move a little bit quickly through this, 'cause we only have an hour, and we have a lot of content to cover. So, ALS is also known as Lou Gehrig's disease. It is

fatal, it's progressive. As I mentioned, it's a neurodegenerative disorder. And it results in profound muscle weakness and wasting of nearly all voluntary muscles, including the muscles needed for respiration, and the number one cause of death for these patients is respiratory failure, because it affects the muscles of the diaphragm, affecting the patient's ability to breathe eventually. Just in the name, Amyotrophic Lateral Sclerosis, there's two main signs of the disease: upper motor neuron and lower motor neuron. And the term, amyotrophic, refers to the lower motor neuron portion of the disease, because that has to with muscle atrophy, and then lateral sclerosis has to do with the upper motor neuron portion of the disease, because that speaks to the degeneration and eventual death of motor neurons in the motor cortex, brain stem, anterior horn cells and the pyramidal tracts, which are all part of central nervous system upper motor neurons.

Patients with ALS face substantial occupational challenges, as many of the activities that bring meaning and purpose to the lives of these individuals, their friends, and family members are substantially altered. All occupations that are characteristic of adults are affected by ALS in some manner, including activities of daily living, instrumental activities of daily living, work, leisure and socialization. The person with ALS may eventually lack the cognitive and physical abilities to adapt to occupational challenges and participate in occupation, or the person may no longer be a match for the occupation, creating occupational dysfunction and loss of identity for persons with ALS. Also, there is just a lack of evidence focusing on interventions whose outcomes are at the level of occupational participation for patients with ALS. In fact, a systematic review done by Erin Foster in 2014 revealed this gap in the OT literature for all neurodegenerative diseases. In terms of occurrences, it's a pretty rare disease. It's 1 to 2 per 100,000 persons worldwide. It's more common in men until the age of 65, and it peaks between 40 and 70, although I've treated patients who were in their twenties when diagnosed with the disease. There's about 5,600 diagnosed yearly. 5 to 10% of the cases are familial ALS.

That means, well, we'll get into the pathophysiology of the disease, many of which are genetically-based. But the SOD1 gene, which I will talk about in a few more slides, is the gene that's associated with the familial form of the disease. And like I said, it's a pretty small percentage, it's only 5 to 10%. And the median survival rate is 32 months from onset of signs and 19 months from diagnosis. Some negative prognostic indicators for patients with the disease include the following. Being older in age at the time of the diagnosis, being female, being diagnosed in a short time after the onset of signs and symptoms, having bulbar onset ALS. Bulbar onset ALS has to do with dysarthria, dysphagia due to degeneration of the motor neurons in the face and the cranial nerves, but also has a higher link to respiratory symptoms from ALS. Also, a negative prognostic indicator is lacking a marital partner, and I think that's just because of not having social support or caregivers available.

Treatment considerations for patients with ALS focus on a multi-disciplinary care that addresses quality of life and symptom management. Okay, so ALS causes weakness to the diaphragm muscle, which I spoke about. Respiratory failure is the most common cause of death, and many patients as they progress through the disease process, eventually come to the decision where they have to either accept mechanical ventilation or go to palliative and hospice care. Now, we're gonna talk a little bit about the signs of ALS. The onset is typically one or a combination of either lower motor neuron onset, upper motor neuron onset, bulbar involvement, cognitive impairments or respiratory involvement. And when we talk about upper and lower motor neurons, I just wanted to point out here that the brain and the spinal cord, so you have the cerebral cortex, the midbrain, the spinal cord, those are all upper motor neurons. And then, when we talk about lower motor neurons, it's really just the nerves that connect the spinal cord to the individual muscles.

Lower motor neuron signs of ALS. So, lower motor neuron signs of ALS have to do with asymmetrical muscle weakness and atrophy. It usually affects the extensor muscles of fingers and wrist first. It also affects the intrinsic muscles of the hands, and typically you'll see more weakness in the thenar muscles than you do in the hypothenar muscles. Dorsiflexion weakness is also a sign of lower motor neuron onsets. So, that's your inability to dorsiflex your ankles. And then, functional impairments from lower motor neuron signs usually are frequent tripping during ambulation, so you might have patients who trip on uneven ground or trip on carpeting, difficulty buttoning shirts, difficulty writing and typing, and difficulty holding or turning a key. Some other lower motor neuron signs are fasciculations. So, fasciculations are small, localized and involuntary muscle contractions. All of us experience fasciculations from time to time. And they're due to a spontaneous depolarization and repolarization which is causing localized muscle twitching. But the fasciculations associated with ALS are pathological in nature and due to damage to the lower motor neuron. And it typically is indicative of the disease advancing. Patients who already have weakened musculature, and they overuse muscles, might experience fasciculations in those muscles and also as the motor neurons in the muscle start to deteriorate. It's sort of a sign that the disease is advancing. Other lower motor neuron signs are cramping and a lack of reflexes or hypo-reflexia. Upper motor neuron signs have to deal with spasticity, and spasticity is velocity-dependent resistance to a stretch.

So, as you're doing passive range of motion to a muscle, you'll feel resistance to that stretch, and this occurs due to legions in the upper motor neuron of the primary motor cortex. This primary motor cortex is responsible for voluntary and purposeful excitation and inhibition of the lower motor neurons, which results in coordinated muscle contractions and functional movement under normal circumstances. But degeneration of the primary motor cortex causes loss of control over the lower motor neurons, and this causes a constant state of contraction, which is spasticity. And spasticity will functionally cause difficulty with ambulation, ADLs and IADLs. We use the Modified

Ashworth scale as a way to measure the degree of spasticity, and it ranges from 0 to 4, where 0 is no increase in muscle tone to 4 is all affected parts are rigid in flexion or extension, sort of seeming like a contracture. But then along the way, there's different grades: 1, 1-plus and 2. And you could look over this in a little bit more detail if you want. After the lecture, you will have these slides. If for some reason, you're unable to do a test on spasticity, you would grade it as a 9 in that limb. Other upper motor neuron signs are hyperreflexia, and this is an increase in sensitivity to deep tendon reflexes. The Babinski sign, having a positive Babinski sign would be a sign of hyperreflexia with an upper motor neuron onset of ALS.

Other signs and symptoms are cognition. So, until recently cognition really wasn't thought of as being part of ALS. But there's a specific type of dementia called frontotemporal dementia, and a lot of patients who have ALS end up also having frontotemporal dementia. And the two diseases, we're gonna talk about it in a few upcoming slides, are so interrelated, pathophysiologically, that some researchers believe that they're part of the same disease process. It's a specific type of dementia that differs from Alzheimer's disease. It's often actually misdiagnosed as Alzheimer's disease despite the clear differences. And frontotemporal dementia occurs, because the neurons in the frontal lobe start to degenerate, and we know that the frontal lobe helps with emotional control, awareness of behavior, recognizing social norms and adjusting to social situations, adjusting your behavior to different social situations. Patients with frontotemporal dementia have behavioral changes, a disregard for social norms and apathy. It's not as much associated with memory loss as Alzheimer's disease, and patients with Alzheimer's disease, for example, can still maintain social relationships, usually well into the disease, at least till the end stages of the disease, but patients with frontotemporal dementia cannot. These patients have a lot of executive function impairments as well because of the degeneration of the frontal lobe. And patients with impaired executive function due to frontotemporal dementia have difficulty processing and interpreting visual and auditory stimuli, organizing information

to form solutions to problems, executing logical solutions and anticipating consequences. Patients with FTD also have been found to have difficulty understanding verbal and written language, as well as having decreased speech fluency.

So, just for example, these are just some of my own examples of patients who have had frontotemporal dementia. There was a gentleman I once treated with ALS that, while driving with his nephew, got stopped at a red light and after the light turned green, he was unable to recognize the green single and associate it with the action of continuing to drive. He said to his nephew, "I do not know what to do next". There was another example where a woman with ALS who was known for being very money conscious. She began spending all of her money on items from infomercials, such as QVC and the Home Shopping Network. And then, all of these items that she was purchasing, they were all items that she would never have purchased in the past and had no current use for. She lost the capacity to reason, self-correct and control her impulses. And then, a third example is a man with ALS who was known for being very warm, friendly and loving, especially towards his family. But he had a complete personality change where he began to be very cold and distant from his friends and family, and he had little insight into his behavior. And when asked about it, could not understand why his friends and family were concerned.

So, those are just some examples of what the behavior pattern for patients with frontotemporal dementia would be like. Other signs and symptoms are, there's bulbar onset ALS. And as I mentioned before, this is caused by deterioration of the cranial nerves. It causes facial weakness, tongue weakness, difficulty swallowing, dysphagia, dysarthria, and an excessive production of saliva and difficulty managing those secretions, otherwise known as sialorrhea. The cognitive profile of ALS. Even in the absence of frontotemporal dementia, there is a cognitive component of ALS. And a meta analysis done in 2016 showed that even in the absence of frontotemporal

dementia, the cognitive profile of ALS is impairment in speech fluency, language, social cognition, executive functions and verbal memory. When we talk about executive functions, we're talking about anticipation, planning, execution and self-monitoring. Other signs and symptoms of cognitive onset ALS is pseudobulbar symptoms, and this is where there's just an inappropriate behavior, like inappropriate laughing and inappropriate crying or what some refer to as emotional incontinence. You might find that just in introducing yourself to a patient, they might start laughing, and then start crying, and then go back to normal behavior, all within a few minutes. Patients who test positive for executive function impairments at baseline have a faster rate of motor function decline and death.

So, the cognitive onset of ALS and cognitive symptoms associated with ALS, are associated with a quicker decline in functional status and usually have a lower life expectancy. Respiratory symptoms with ALS are dyspnea on exertion or difficulty breathing when lying flat, weak or ineffective cough, you'll see increased use of auxiliary musculature when attempting to breathe. And then, because of oxygen starvation and a decreased oxygen saturation, patients with respiratory symptoms will have daytime sleepiness, decreased concentration and headaches. But ALS usually does not affect bowel and bladder function, internal organs, sexual function, just from the standpoint of sexual arousal. Obviously, with the physical, sensory motor function decline with ALS, engaging in sexual activity might be difficult from that standpoint, but it doesn't affect sexual arousal or anything like that. Sensation is usually not affected, because sensation is related to the posterior horn of the spinal cord, which is usually not affected by ALS. Also, eye musculature and eye movements are typically not affected, which is good for communication software devices, because you could use eye gaze software to be able to communicate and use a computer, and operate items around your house like lights and things like that. All right, now we're gonna get into a little bit of the pathophysiology of ALS. The TDP-43, which stands for Transactive Response DNA Binding Protein 43, it's a protein that is normally present in the cellular

body of neurons. It plays an important role in neuron cellular functioning, because it binds DNA and RNA in neurons, and as I said, it's normally present within the cell body. But with ALS NFTD, there's an excessive accumulation of TDP-43 found in the intracellular portion of motor neuron tracts in patients with ALS and in the intracellular portion of neurons comprising the frontotemporal lobe in patients with frontotemporal dementia.

So, this graph sort of summarizes that a little bit better. I'm gonna try to do this a little bit better this time. So, on this patient on the left, this is a patient with ALS. And you'll see the cerebral cortex and the primary motor cortex there in the red represents the TDP-43 excessive accumulation in the intracellular portion, and you'll see it in the primary motor cortex and the spinal cord. So, that's a patient with classic ALS. This control subject has TDP-43 present, but it's all contained within the neurons, and you don't see any in the motor cortex or the spinal cord. And then, if you come over here to this brain, you see the TDP-43 in the frontal lobe, and this is a patient with frontotemporal dementia. If you had a patient with both ALS and frontotemporal dementia, you would see the TDP-43 in the motor cortex, the spinal cord and the frontal lobes. So, there is a pathophysiological link between ALS and FTD. The molecular and genetic link between these two diseases have led some researchers to believe that these are actually the same disease process, and they should be considered the same disease. Some other causes for ALS is C9ORF72. It's a gene located on chromosome 9. It's associated with a protein binding for axonal and dendrite growth. So, this gene creates this protein for axonal and dendrite growth. And it's normally found in the cytoplasm in the presynaptic terminals of neurons. But the mutations to this gene, so that there's a lack then of this binding protein, has been linked to ALS. And patients who have a mutation to this gene have a lower age of onset for ALS, impaired cognition, impaired behavior, abnormal changes in the gray matter in their frontal cortex, and a reduced time of survival.

Then, there's the familial form of ALS. Again, this only accounts for 5 to 10% of cases. But the SOD1 gene, the SOD1 gene encodes the enzyme, SOD1, which is superoxide dismutase 1. And it's usually present on chromosome 21, and it's actually this enzyme that this gene creates as an antioxidant that regulates superoxide. The superoxide is a byproduct of oxygen metabolism, that if left unregulated, will cause cell damage. And then, of course, if you don't have this antioxidant, mutations to the SOD1 gene have been linked with ALS. Some other theories on the cause of sporadic form of ALS, and by sporadic I mean it's not familial, is an excessive amount of glutamate, which is a normal neurotransmitter associated with memory and learning, some sort of neuro-inflammatory process, which I think is more stress-induced. In my experience, a lot of veterans, especially veterans who have served in foreign wars, and actually police officers, in my experience with treating patients with ALS, have developed the disease. And I think that comes along with, because of the stress of the job and the neuro-inflammatory process.

Other causes could be environmental, head trauma. We all know about football players and soccer players being prone to having ALS. Also, excessive physical activity, a lot of marathon runners and other athletes that are engaged in a lot of physical activity are a little more prone to having the disease. And of course, we talked about the genetic predisposition. So, what does a typical person with ALS look like? It's actually a trick question, because there really is no standard presentation. They use the El Escorial Criteria to diagnose ALS, and this sort of just goes through it. You can read through it. It moves from suspected ALS to definitive ALS, And it all really depends on the amount of lower motor neuron signs, and upper motor neuron signs, and the progression of the disease. The one thing, whether you have suspected, possible, probable or definite ALS, is the signs dealing with upper motor neuron and lower motor neuron signs cannot be explained in any other way. There can't be any electrophysiological or neuro-imaging evidence for any other disease process. And that's how they diagnose the disease.

You might notice on here that there isn't anything around cognitive function as well. That's one thing that's sort of left out from the El Escorial Criteria. So, treatment and medications. What drugs are available to treat ALS? The number one drug that patients are prescribed is Rilutek, and this has to do with glutamate. Remember we said that there was a theory that excessive glutamate was one of the causes. Riluzole, or Rilutek, it just reduces the amount of glutamate in the system. It's approved by the FDA for ALS treatment, but it really only increases survival by two to three months. And I don't know if anything has changed, but at least back in 2012, and before that, it was not typically approved by insurance and was expensive. There were many patients who decided not to take it, because the benefits of survival for two to three months they just didn't feel was worth it. There's another drug that's given orphan drug status by the FDA, meaning that it's not fully approved yet, but you can prescribe it for ALS and that's NPT520. This is a drug that penetrates the blood-brain barrier. This has to do with the SOD1 gene, so it decreases the neurotoxic misfolded proteins that come in the absence of superoxide dismutase 1. That antioxidant produced by the SOD1 gene, it takes care of those misfolded proteins that result because of excessive oxidation.

And then, there's Tiglutik which is really just the liquid form of Rilutek, or Riluzole, and it's a liquid form that's administered with an oral dosing syringe for patients who can't tolerate the tablet form. And then, there's Radicava which removes free radicals, provides neuro protection, and it basically takes the place of the SOD1 gene by providing some antioxidant protection in the absence of superoxide dismutase 1. You might notice that a lot of these drugs really focus on that SOD1 gene, and it's really only 5 to 10% of ALS cases. But I think it's because the SOD1 gene was the first gene discovered as one of the causes for ALS. So, it's been around longer, so there's been more research on it. Hence, the reason why there's more of a pharmaceutical response to it. Hopefully, in the upcoming years, there will be more that look at the TDP-43 aspect of ALS.

So, what are the goals for PT and OT when treating patients with ALS? And it's really activity/exercise modification, stretching, range of motion, trying to reduce pain, normalize tone, prescribe and utilization of appropriate assistive devices, patient and family education and a lot of focus on reducing caregiver burden. I wanted to talk a little bit about exercise, only from the standpoint that, obviously, if you're gonna have a disease that's going to reduce the strength of all voluntary muscles, the patient and you are gonna wonder should I be prescribing exercise? Because we live in a society where we think that you have to stay active, and it's use-it-or-lose-it type of mentality. And it's a little controversial in patients with ALS, because you do have to remember that they do have a finite amount of energy and muscle strength during the day, and if they overuse muscles or over-engage in activity, they're gonna be fatigued. And that might make it that they can't engage in other activities later in the day, after really trying to perform and exercise or engaging in an excessive amount of activity earlier in the day. But it does have it's place, and obviously, exercise is an activity that some people, like for me, for example, I like to run and lift weights, and it's an important part of my life. And a lot of patients who have ALS, they don't wanna lose that. So, it does have it's role. But there was this one study that showed that there is some benefits to at least staying active. And I know we're occupational therapists, and we're not focusing all on just prescribing exercise, and I'm not suggesting you do that, but you could take these considerations when you're thinking about engagement in activities and purposeful and meaningful activities. Because the exercise can just as easily be replaced by engaging in meaningful and purposeful activities, which may increase the quality of life.

So, this was a randomized control study, and it only had participants, I wanna point out, who had a manual muscle test of 3 out of 5 or higher, so these weren't highly advanced ALS subjects. And the intervention groups, they were in this program for six months, and they were split up into three groups. They either got active exercise performed with a physical therapist in a clinic plus cyclogometer activity, which was 20

minutes of upper and lower extremity exercise activity in a seated position. So, they either were put in that group, or a group that just got active exercise without the cyclogometer, or a group that just got passive range of motion protocols. And what they looked at was: what was the ALS Functional Rating Scale? The ALS Functional Rating Scale rates bulbar signs, upper limb function, lower limb function, and respiratory function. It consists of 12 questions, centralizing on various functional activities, which is including speech, salivation, handwriting, dyspnea, walking or dressing. Although, I do wanna point out that the ALS Functional Rating Scale does not exclusively look at executive function or behavior. So, I looked at their score on that, their forced vital capacity, how much air the subject was able to breathe into their lungs and then expel out of their lungs. And then, their perception of quality of life on the McGill Quality of Life Questionnaire. All participants were measured prior to treatment and then monthly until the end of the exercise or non-exercise groups, and then again six months after treatment. And what they ended up finding is that the patients who had the active exercise, especially the group that had the cyclogometer components, who had performed better on the ALS Functional Rating Scale throughout. And they even were better at the six-month follow-up, the exercise group. However, there was no significant difference in quality of life, and no significant increase in survival were found.

So, exercise does have its role. But as I said, it can be controversial and is potentially detrimental in muscles that are exhibiting weakness. These patients did have weakness, because they were 3 out of 5 on a manual muscle test. But I can tell you from experience, I've had patients, and for some reason it was males, who had some shoulder flexion, shoulder abduction, external rotation. Let's say, maybe they were 3-plus to 4-minus out of 5 on a manual muscle test, and 3-plus to 4-minus out of 5 on elbow flexion. And upon evaluation, I gave them my spiel about really trying to pick activities and exercises that are meaningful for them and don't overdo it. But these gentlemen decided that, well, they wanted to continue going to the gym. And this one

individual I'm thinking back to really did a lot of exercise in between our sessions. And he was going to, basically lifting weights to the point of failure, and within a one to two-week time span, went from a 3-plus to 4-minus out of 5 to a complete 0 out of 5 on a manual muscle test. I've seen this before.

So, to that second bullet point, that it can't possibly be detrimental, I know from experience that it can be. So, you just wanna be careful when you do have a patient who wants to engage in exercise or even excessive activity and have them stop if cramps or fasciculations are occurring. Also, avoid eccentric contractions. They've been shown in the literature to be a little bit more damaging to muscles than concentric contractions. And also, avoiding high resistance and high repetitions, just monitor their fatigue level and do not give resistance exercise to already weakened musculature as I spoke about in that example. The best thing they could do is stretch. You wanna incorporate a lot of stretching to normalize tone and maintain range of motion. These are just some guidelines for doing stretches. Definitely wanna involve the caregivers, because that stretching in itself is an activity and as the disease progresses, the patient might not be able to effectively stretch. So, we cannot increase the muscle strength, but we can maintain muscle length with stretching. It also helps with spasticity management.

All right, so now we're gonna transition to talking about some OT theories to help guide us when we are treating patients with ALS. And this first model that we're gonna talk about is the ecological model of human performance. This is one of my favorite models or theories in occupational therapy, because I think it really captures what we do as occupational therapists, and I also think it provides an avenue for us to describe what we do to other professionals. I do encourage you to read more about this. 'Cause using the language associated with the EHP might help you communicate to physical therapists, nurses, physicians, social workers, about just what occupational therapy is about and have them develop a greater understanding about occupational therapy.

This is a patient-centered approach that considers the person's personal interests, values, experiences, and how they relate to person's temporal, physical, social and cultural context, in regards to performance.

We talk about ecology. This is the study of the relationship between organisms and their environment, which provides an excellent context to examine the performance range for persons with ALS as they cope with changing and diminishing physical and cognitive skills, but still have a continued relationship with their environment. So, some key constructs of the EHP. There are person factors. We all have person factors. These are sort of abilities that we possess, they reside within us. And these are our sensory motor skills, our cognitive skills, our psychological abilities, personal interests, values and experiences. And then, we are surrounded, if you go down to the third bullet point, we are surrounded by a context. And these are things we don't necessarily have full control over. The contexts are sometimes temporal factors, which are time-bound, like age. I mean, we're all gonna age, we can't control that. Life cycle, health status and expectations. There are physical factors that surround us. It's the objects and the elements that surround us. Social factors such as the relationships and norms, and your relationships and norms change depending on who you're socializing with. You might have a different relationship with one person that's more formal to one that's more cordial, and you can have a completely different relationship with different sets of people.

Also, the cultural/environmental factors that surround us, the beliefs and values of the society that we live in. Not only just even in our immediate environment. For example, there might be different cultural and environmental factors for someone who lives in New York City compared to someone who lives in North Dakota. And also, different cultural and environmental factors from someone who lives in the United States to someone who lives in Iran. So, the context surrounds us no matter where we go, and it could change depending on where we go. And it's how we interact with our context

and how we use our person factors to interact with those contextual factors to form a transaction for which we have complete tasks. And a task is an objective set of behaviors needed to engage in performance to reach a goal, and the performance refers to the person's engagement in tasks within the context.

So, some core assumptions of the EHP. As I alluded to, it is impossible to examine a person outside of the context in which they live their lives. And performance occurs, as I said, as the person engages in a transaction with his or her context to engage in tasks and combinations of tasks. So, the more tasks you're completing, these tasks can be grouped together to make up roles and occupations. Context can enable and promote, as well as obstruct and inhibit an individual's performance and performance range. For example, a person with ALS. If they, under normal circumstances, before they had the disease, they were interacting with their context very well. They were independent. And their context, let's just say, their physical environment of their house, a two-story house, they were fine, they were independent going up and down the stairs. But now, with a decrease in sensory motor function, living in a two-story house, that context now no longer supports them in independence. And it's now hindering and inhibiting performance. And the performance range is determined between the person and the context: the more effective that transaction, the greater the performance range. And independence occurs when a person's wants and needs are fulfilled. And also any assistive device or support that's provided to the patient, the EHP model views this just as a tool to achieve independence.

So, there's no concerns with using assistive devices or activity modification with the EHP. And this model right here, you'll see in that center circle, you have the person, containing the person factors, surrounded by the context, and that arrow is the transaction with the context, and you could see in this case, because that person is transacting so well with the context that they're able to complete a large number of tasks, which if grouped together make up occupations. If that transaction did not work

very well, then the performance range would be much more narrow. Another treatment approach that I wanna talk about is the neurofunctional approach. So, the neurofunctional approach was developed in the late 1980s and early 1990s by Gordon Muir Giles and Jo Clark-Wilson as a method of increasing occupational performance for patients with traumatic brain injury. It's best used for patients with ALS as a framework for assessment and intervention and to actually targeting occupational performance and not cognitive processes.

As occupational therapists, we often defer to neuropsychological approaches, and I've done this myself. For some reason, it's very prevalent for us to use cognitive rehabilitation which focuses on retraining cognitive processes, such as short-term memory, attention, orientation and basic problem solving. The NFA differs fundamentally from the cognitive rehabilitation approach by directly targeting occupational performance in real-world activities and not attempting to retrain these cognitive processes. We know in most literature it shows that you can't train short-term memory as if it's some sort of muscle that can be improved that will then be generalized to all functional components. You can't separate out individual cognitive processes, because they're all interrelated, and they all directly relate to function. The NFA also differs from other neuropsychological approaches that use metacognitive strategies. And metacognitive strategies are your awareness of your behavior and developing strategies to become more aware of your behavior and then changing your behavior in the moment. And the reason why the NFA is different is because evidence suggests that these techniques just are not effective for persons with such a severe lack of insight and behavioral impairment, especially those individuals who have ALS who might have this lack of insight.

For example, you might remember CO-OP. It's another OT theory, the Cognitive Orientation to daily Occupational Performance. This is an example of a theory and a method that uses metacognitive strategies based on awareness and insight. It uses the

goal-plan-do-check strategy. The NFA is completely different than that, it's a more prescriptive approach that directly addresses the functional impairment at the level of occupation. It uses a precise method of performing a task to where automaticity is achieved. And what we mean by automaticity is that, automaticity is a state where behavior is mastered to the point where the cognitive demand and the cognitive energy spent on performing that activity is completely removed, because it's just an automatic behavior. And the biggest example would be with driving. If you remember when you were learning to drive, you had to attend to every decision you were making on the road. But now, under normal circumstances, unless it's an emergent situation, you don't even have to attend to driving. You can keep the car on the road, turn left, turn right, apply the brake, and you're not even thinking about it.

I'm just gonna go forward here just a little bit. So, these are just some approaches that you would use in the NFA approach: overlearning, errorless learning, and chaining techniques. Overlearning would be deliberate over-practicing of a task past a set criterion. Errorless learning are tasks that are taught in a manner designed to eliminate the number of errors made during the learning phase. And then, chaining learning would be learning whole, functional tasks by utilizing a step-by-step process to teach sequential steps of a task until completion. And chaining can be forward chaining, backward chaining, or whole task training. When working with ALS patients on performing ADLs, let's say, in a certain manner with activity modification, sitting versus standing, making use of assistive devices. This is the type of approach you would use with patients who have ALS, because you want them to do it in a specific fashion completely altering the method that they were using yet before, with the goal being to make them more efficient and effective at performing this task. And this is just a summary.

This is something I developed in my doctorate program. And it's called the Ecological Approach to Optimizing Occupational Participation for persons with Neuromuscular

Disease or the EOP-NMD. You won't find this in the literature, it's just something, I was just working on it. It takes components of the EHP and the NFA, but also adds my own sort of experience with working with patients who have ALS. And this is how I view working with patients who have ALS. Instead of having the person surrounded by the context, I sort of envisioned having the person and the context as two separate circles that overlap, and the degree of overlap of the two circles, in the middle, you'll see occupational participation. And we as occupational therapists, in some manner, may want to push the two circles together to increase the degree of participation for patients, or in some cases, we might wanna pull them apart. And the reason you might wanna pull them apart, is because the activity is just not that meaningful for the patient or maybe on certain days there's some activities that are more important than others, like, for example, if you have a patient who has a family outing that they really want to attend.

Let's say it's a birthday party, a grandchild's birthday party, that they really want to attend later in the day, you don't wanna use up all of their energy at the beginning of the day performing ADL. So, you would want them to have more assistance earlier on in the day. So, you'd want to pull the two circles apart and limit the amount of participation for ADLs, so that they could attend the birthday party. But then, maybe on some other days, there's some other activities that are more important. And maybe on some days the ADLs are important to the patient, and you'd wanna push them, the two circles, together. And you would do this through a therapeutic partnership, the relationship with the patient should be based on trust and understanding, and using motivational interviewing techniques, which call for a relationship based on acceptance, empathetic understanding, relationship listening skills, and care taken not to convey judgment or confront the patient in any manner. We as therapists, when working with these patients, should always maintain a therapeutic partnership. You wanna enhance cognition to reestablish or maintain habits or routines through practice.

That's your functional cognition, that's using the NFA and some elements of the ecological model of human performance. You wanna collaborate with caregivers and interprofessional staff, reducing caregiver burden whenever possible, get having strategies so that there's extra support for patients and also advocate to patients to other interprofessional staff. You wanna work with the patient in the natural context. The EOP-NMD is not designed to be working with patients in a clinic or a simulated environment. This has to be performed within the natural context. And then, much like the ecological model of human performance where you're trying to better match the person's abilities to the context through activity modification, through use of assistive devices, just being aware of their energy levels, and you would better match the context to the person's abilities.

So, those are the five essential elements of the EOP-NMD to either bring together and enhance occupational participation, or in some cases, you might wanna, as I mentioned, decrease occupational participation. All right, so we're gonna spend the remainder of this course going through just some common interventions for patients with ALS. The first one we'll just look at is mobility, functional mobility. And I'm just gonna be giving some general interventions. So, a cane obviously is good for someone who lacks balance and has lower extremity weakness, but I would recommend using a single point cane versus a quad cane just because it's easier to lift, it doesn't add as much weight. Obviously, we talked about lower motor neuron onset of the disease and having a lack of dorsiflexion. An AFO is sometimes required. I would recommend a carbon fiber AFO, because it's just lower in weight and less bulky than some of the other AFOs you might come across. Rolling walkers are recommended, but the ones with handbrakes and that you would sit down on, otherwise called a rollator, aren't always recommended. Just be careful, if you are prescribing a rollator, that the patient has adequate hand strength, trunk strength, and also just the awareness and ability to be able to utilize the brakes and make sure that the brakes are on before they sit down and that they're able to operate it independently. Wheelchairs obviously are great. I

would recommend a companion wheelchair for any of the patients, just because it's a tool to allow you to go further into the community. And you could take it with you in a car, easily in and out of a trunk. Caregivers can push the patient in the companion wheelchair, and it does not have to be all-or-nothing. They don't have to exclusively use the companion wheelchair, it's just there when they need it.

So, for example, you might have a patient that can walk a certain distance, but if they go further than, I don't know, I'm just throwing out there, like 500 feet, or they're walking for more than 10 minutes, they might need to sit down. So, a companion wheelchair would be great for those patients. For power wheelchairs, just make sure that you are recommending the right power wheelchair, and you're not using your insurance benefit on something like a scooter or a power wheelchair that does not have tilt in space or other positioning that patients might need, like lumbar positioning and support. You would be making recommendations as an occupational therapist for the type of controls on that chair, where those controls are located, and also teaching the patient how to use the chair. Even in the absence of being able to use a power wheelchair in the community, even if they just use it in their house for positioning, it's a great tool to use, because it does reduce caregiver burden and provide some level of control to the patient. If they wanna move locations and go to a different room in the house, they could do that on their own. If they wanna change positions like raise their legs or decrease the amount of hip flexion that they're in, they could do that on their own independently and do not have to rely on their caregiver. It really, if used properly, can increase quality of life by decreasing pain, just increasing comfort, and just increasing independence in the home, and if they use it outside in the community, it's even better.

And we usually recommend it earlier on in the disease process, because if they do get fitted for one, it could take three to four months for them to get their power wheelchair, and you want them to get the most out of it. I mean, you're only looking at a 19-month

span where patients will be living with the diagnosis of ALS. Just some mobility continued here. Family training on the right techniques to reduce caregiver burden. We don't want caregivers getting hurt while transferring patients, that's a big thing. Knowing when to limit or avoid ambulation. Again, it's not an all-or-nothing, but maybe you wanna give them some cues that you can ambulate and walk on your own, but you don't wanna go for an extended period of time and having the patient become a little bit more aware of when they're starting to get fatigued, so they don't overexert themselves.

Also, use of a Hoyer lift. It's a great tool to have on hand. You do not have to exclusively use the Hoyer lift. It could be used when patients are particularly fatigued or on days when they're just not feeling well. And on other days, maybe they transfer either independently or with the help of a caregiver but having it on hand is helpful. And another common myth about the Hoyer lift is that it's big, it's bulky, and it's difficult to use, I can assure you that with the right practice it is easy to use it in even the tightest of environments. So, I do recommend exploring that with patients. And with a lot of these tools, it's not always easy to get patients to accept them, so it's not always a one-time conversation. Remember that therapeutic alliance and trust. Sometimes it just might take a few conversations to get patients to come around to accepting a lot of these devices as tools. Just like with the ecological model of human performance and the NFA, these are considered tools to just enhance independence and as a method to better match the patient's abilities with the context. Some more information on equipment prescription, I think we kinda went over this. If you wanna look them up, like what a pivot disc is, that's a great tool for stand pivot transfers. Patients who could do sit-to-stand and have some strength in their hips and trunk, you could use this to help turn them to sit on a chair.

The one thing I do wanna point out, transfer boards, I don't always recommend them. It takes a lot of energy, and it could be very difficult, especially for patients that have

advanced weakness, like in their upper extremities, trunk and lower extremities. I would recommend at that point, if you're thinking that they need to use a transfer board, I would probably go to either a more dependent stand pivot transfer, maybe try a pivot disc, or go straight to a Hoyer lift in that case.

Activity adaptation, we talked about energy conservation, any time you could sit versus standing, especially for things throughout the day, 'cause it all builds upon itself. The more time you spend, these patients perform activities while standing, the more fatigued they'll get later in the day. So, you always wanna incorporate energy conservation throughout the day. One thing is driving, and you have to consider all aspects of driving. It's not just about keeping the car on the road and having the ability to turn the key or shift gears, but you also have to think about decision making and that example with that patient who was stopped at the red light for frontotemporal dementia. And also, where it is that they're going, because eventually wherever they're going, they're going to have to get out of the car and then ambulate. And if they're going in independently, you could have a case where the patient could reasonably drive a car and get from point A to point B, but are they safely going to be able to get from the parking lot to wherever it is that they need to go and navigate uneven ground and walk those distances.

So, I often have a conversation about thinking about alternate forms of transportation prior to it becoming an issue. And it's difficult, it's difficult to have these conversations, because driving just represents so much independence for these patients. We have had patients who had the resources to modify their car, which is great, but again, you have to remember about the transportation and all those other factors that I just spoke about, in terms of walking across parking lots and things like that. All right, ADL prescription. Reachers, button hooks, universal cuffs, they're all great. Sock donners not always useful, just because of the amount of fine motor skills and hand function that it takes to load the sock onto the sock donner and utilize it. I usually try first to do

a leg-crossover technique or some alternate method before I go to a sock donner for these patients. Just an example of a button hook with a built-up handle using a cylindrical grasp. It's a great way to button shirt buttons. Another method is just to keep the buttons buttoned and do just a one-handed dressing technique, or dress your arms first, and pull the shirt up over your head, thread your head in last technique. That's another way to get around shirts with buttons. Or, just avoid shirts with buttons. This often comes up with men who are still working that have to wear dress shirts to work or want to wear dress shirts to work. I usually work with them on some method so that they're able to still dress, look presentable, feel professional, but not overexert themselves with dressing. So, there's that aspect of the role that comes along with dressing that we have to consider as OTs.

This is just for feeding. Feeding is very important and nutrition intake is very important for these patients. Oftentimes, because of advanced dysphagia, they have to face the decision of whether or not to have a PEG tube placed. And nutrition intake is really correlated with not only quality of life, but length of life and lifespan for patients with ALS. They actually want them to take in a lot of nutrition. So, we as OTs really have to work on maintaining independence with self-feeding. A tripod grasp is gonna be a lot more difficult to use than something with, like this, with a cylindrical grasp. This is a great setup for a patient for self-feeding. You can see her arm is flexed at the shoulder at almost 90 degrees, and then it's supported at the elbow, so she doesn't have to abduct and flex her shoulder and then she only has to use a flexion and extension of her elbow using a built-up handle spoon with the food raised, so it's easier to bring the food to her mouth. Hand weakness is very prevalent in patients with ALS. As I mentioned, hypothenar weakness, thenar weakness, you'll see ulnar drift, flexion and extension contractures. I usually recommend using resting hand splints, wearing them at night if it's bilateral hand weakness that we're worried about. And it's a hindrance for them to wear hand splints, usually have them wear the right-handed one one night and then the left hand the other night, and if it's too much to do that, maybe two to three

hours during the day, just to get some good positioning and maintain the joint integrity. Because they're hands could become very painful and deformed as well.

Also, shoulder subluxation is a big part of this disease. I usually recommend the GivMohr sling. You can actually google that, G-I-V-M-O-H-R. It's a good sling that helps hold the head of the humerus in the shoulder. Cervical support, this is a Headmaster, and there might be other forms of this type of soft collar. It allows for some posterior flexion and anterior flexion and rotation and lateral flexion of the head. It allows them to be able to chew and communicate. It doesn't provide a lot of compression around the neck, interfering with respiration. So, this is usually the cervical support that I recommend first. It's rigid along the collarbone and rigid along the chin, although it's like a little soft pouch that you could rest your chin on. It can become soiled, and it can wear out over time, so they might need a new one after a while. If someone really has advanced weakness in their cervical spine and upper thoracic area, you might wanna recommend a Miami J or something else that's a little bit more comprehensive than this, but this is where I usually start. Computer use, just refer back to your ergonomic setup. Voice typing software, as I mentioned, eye gaze software and on-screen keyboarding are great for patients with ALS. You could work with them on proper positioning and use of the software, so that they're able to maintain their role as a worker and communicate, and use social media, and interact with patients.

Remember, with OT it's not all about ADLs, we should be really working on more meaningful activities for patients, because quality of life is really going to be enhanced if they can still interact with friends and colleagues and feel like they're engaging in purposeful activities. So, an ergonomic setup for computer use is a big part of occupational therapy treatment. This is just some more information about wheelchair prescription, we talked about that. ALS is not a painless disease. So, despite what you may have heard. So, as I talked about stretching, positioning, other modalities like ice

and heat can help. There is medication that you can advocate to the physician to prescribe for the patient, but remember that does have side effects, and trying to find the pain and addressing any pain issues that might come from weakness or joint instability before it becomes an issue is important for OTs.

So, just be proactive with addressing pain. Just some information on dysarthria. You can look through these slides just for some information on patients who have difficulty speaking, and you could use this for caregivers to educate them on how they can better communicate with their loved one. Same thing with dysphagia. A lot of things that I think we already know, and we could learn from our speech therapy colleagues as well. Shortness of breath. Patients often end up having to go on non-invasive positive pressure ventilation, or BiPAP, prior to going to mechanical ventilation. So, it's not about oxygen saturation. Their lungs are fine in terms of getting oxygen into the blood, it's the fact that they can't take air in. So, that's why we recommend that they use BiPAP, especially at nighttime, which will help them feel better during the day. They're not gonna have that sleepiness and sort of brain fog because of a decreased saturation in oxygen. So, a lot of patients will wear the BiPAP machine at night and even for portions during the day, before they make the decision to go to mechanical ventilation. I do have to say that most patients in my experience choose not to go on mechanical ventilation and turn to hospice instead. It's mostly just because there's locked-in syndrome that comes with, and the expense of care, and just the burden on their family, 'cause they can't really interact and locked-in syndrome really has to do with the loss of being not able to communicate and then being hooked up to a device that's essentially just keeping you alive. Just some information on managing secretions.

Psychosocial factors, I talked about that. And I really think that that, being able to engage as much as they can with family, caregivers and colleagues from a psychosocial standpoint is very important, really trying to address depression,

insomnia. And then, there's other things that come along with it, like financial stress, access to nutrition, utilities and bills, really partnering with social work and social services and talking to patients about this. So, don't just focus on ability to function with ADLs and IADLs, really develop that therapeutic relationship with your patient so that you could start to address some of these psychosocial factors that come up with the disease. 'Cause this is really gonna be where it's at with increasing quality of life. Just some other resources from ALS Association, you should look up your local ALS association and partner with them. They have a lot of these resources available. And that actually concludes our lecture. So, I'm sorry if I went really fast. As you could see, it was a lot of material to cover, but we're just at about 60 minutes. And I'm hoping you're all able to stay on, we have some room for questions, and I think Fawn was going to read them off. Or, I could see them right here actually.

- Yes, thank you so much, Scott. Yeah, thank you so much. I'll go ahead and read some for you here. So, Nadia, or Nadia, is asking "Why is being female a negative prognostic factor, especially since it's more common in males? Do females demonstrate quicker degeneration?"

- [Scott Rushanan] That is a very a good question. And to be honest with you, I don't really know the answer to that. I'm thinking maybe it's because, going back to a lack of a marital partner, and that females live longer than males. So, when they are diagnosed with the disease, they're spouses, they might be widowed at the time, and then they might have less support when they develop the disease. That's just one working theory I have right now. But that is a very, very good question, and I wish I would've dove deeper into that. You have me thinking about it now too. So, I think after this I'm going to look that up as to why that is. Sorry, Nadia, if I'm unable to fully answer that. But that's my working theory.

- [Fawn Carlson] Okay, thanks. She has another question: in what kind of case would you be unable to test using the Modified Ashworth scale?

- [Scott Rushanan] I'm thinking if it's too painful for the patient to have their joints moved. Remember, when you're testing for spasticity, it's velocity-dependent. So, you might be, let's say, if you're gonna look at, I'm just gonna use something very basic here, like spasticity in the bicep muscles. And you might have to move the patient's shoulder, but they have a subluxed shoulder, and it's painful. So, you don't really wanna move that joint anywhere near an aggressive fashion. So, you wouldn't test it. So, you would score that as a 9. Or, at the time, the patient was just put into a position that they were very comfortable in, and you didn't wanna disturb that. And maybe, you were seeing them in a clinic, or even in their own home, and at the time, you just could not assess it, you could score it as a 9. Those would be some of the reasons.

- [Fawn Carlson] Okay, Susanne is asking, "How many people are diagnosed with ALS yearly?"

- [Scott Rushanan] I believe that was on one of the slides. Was it 5,600 or something that was on one of the earlier slides? You'd have to look on there, but I believe it's on there. Is that the right number?

- [Fawn Carlson] Yes, that's correct.

- [Scott Rushanan] Okay.

- [Fawn Carlson] And we have another question. I'm not quite sure what she's asking. In your summary of theory, those of us with chronic pain and auto-immune disease, we call that spoon theory?

- [Scott Rushanan] Maybe, she's talking about the ecological approach to optimizing occupational participation? I don't know, Annette, are you talking about the way of the two circles coming together, in maybe I borrowed some aspect of spoon therapy? The way I was talking about the person and the context coming together?

- [Fawn Carlson] Yes, she's saying yes. Okay, and do ALS and dementia have similar survival rates?

- [Scott Rushanan] Is the question: if you have ALS with dementia, like frontotemporal dementia, or just ALS in general? If you have ALS with frontotemporal dementia, your survival rate's gonna be much lower than a patient who just has ALS. And I've had patients... I know I spend a lot of time on the cognitive signs of ALS, but there are patients who just really have upper motor neuron or lower motor neuron signs and really not a lot of cognitive involvement. And they do tend to live longer. I remember I was saying in this lecture 19 months on average is about how long your lifespan is after you're diagnosed. But I'm even remembering one individual who was with us at the clinic for many years. In fact, I think he was there shortly after I arrived in 2004 and when I went over to Home Health as a director, he was still going to the clinic. So, he lived quite a long time. I know he had a few years back passed. I don't know if that's what you're asking? If you're talking about Alzheimer's disease, patients with Alzheimer's disease will live longer than patients who have ALS and frontotemporal dementia.

- [Fawn Carlson] I think that did answer her question. And then, Gabriel is asking, I believe he's saying, "The zip-control wheelchair is contraindicated since they suffer from respiratory problems."

- [Scott Rushanan] The zip-control wheelchair, what is the... Oh, is this with breathing? Like a power wheelchair where you would control it with breathing? Yes, most likely. I

mean, look, anything's possible. But even with that individual that I was just talking about, for example, who lived quite a few years with ALS, and he mostly just had lower motor neuron signs and even not a lot of spasticity, it was just a lot of muscle wasting and becoming weaker, mostly stayed with lower motor neuron signs. Probably, he could have operated a power wheelchair with breathing. I know that they're probably pretty sensitive with the sip-and-puff type of power wheelchairs, but even in the later stages I know he would have had difficulty operating that.

- [Fawn Carlson] Yes, he did mean-

- [Scott Rushanan] Usually Head Array usually works pretty well. There's usually some movement in the cervical spine you can use to control the power wheelchair.

- [Fawn Carlson] Right, I do believe he meant sip-and-puff. I don't see any more questions coming in. So, I think that will conclude for today. Thank you, Scott, for a great time. And all your answers to our questions.

- [Scott Rushanan] Thank you. I hope you all enjoyed it. I know we moved kind of fast there, but I hope you all enjoyed it, and I encourage you look through the slides. If you have any other questions or anything comes up, don't hesitate to ever reach out to me. My information is on there. And again, I enjoyed providing this so thank you.

- [Fawn Carlson] Thank you so much. I hope everyone has a great rest of the day. You can join us again on Continued and OccupationalTherapy.com. Thanks, everyone!

- [Scott Rushanan] Thank you.