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Treatment of Patients with Amyotrophic Lateral Sclerosis
in the Home Health Setting
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- [Calista] I'd like to introduce Scott Rushanan today. Scott is the current director of rehabilitation services and the co-director of Penn Medicine's Home Health agency. Scott previously spent eight years as the lead occupational therapist at Penn Medicine ALS clinic, and Scott is currently pursuing his doctorate in occupational therapy at Columbia University, focusing on cognitive impairments for patients with neuromotor disease. So Scott, we are so pleased to have you here with us today, and at this time, I'm gonna turn the microphone over to you.

- [Scott] Thank you, Calista. Good afternoon, everyone. I'm very excited to talk to you all today about physical therapy interventions for patients with ALS. As Calista had mentioned, I had spent eight years as the lead occupational therapist at Penn Medicine's ALS clinic. This clinic, it occurred every Thursday, for patients who were diagnosed with ALS, and the rotation was about three months, every three months patients would come in for a treatment, and they could come at a higher frequency or a lower frequency, depending on the progression of their disease or what signs or symptoms that they were having. And at the clinic, these patients would receive physical therapy, occupational therapy, and speech therapy. Usually the physical and occupational therapists would come in together and the speech therapist was teamed up with a nutritionist. The patient would also see a nurse, a mental health nurse, neurologist, the pulmonologist, and a social worker.

So it was sort of a one-stop shop to receive all of these services and have this multi-disciplinary team, you know, address any signs or symptoms that the patient may be having due to their ALS. And despite the fact that ALS is a progressive and fatal neuromuscular disorder, I personally, this was a very rewarding experience. The eight years that I worked at this clinic was were some of the best of my professional career, only in that it really, despite it being fatal, there was a lot of good that came out of this team working with these patients and I think we did, there's a lot of good going out into just improving quality of life for the patients and for their family members as they cope

with this devastating disorder. And just from a professional level, when I first started there, I had maybe five years of experience as an occupational therapist but these eight years really taught me a lot about interdisciplinary teamwork towards treating symptoms or signs and functional deficits for patients with a very, very devastating disease, and I've applied a lot of the learnings that I've had, from working at the clinic, to other aspects of my career in even other disorders that are completely unrelated to ALS and not even neurological in nature. So our learning objectives for today, after this course, all of the participants, you'll be able to describe at least three signs of ALS that result in a loss of physical and cognitive function associated with the disease, identify at least one environmental, social, and personal factor that may impact function and participation in patients with ALS, and then we're actually gonna go over two theoretical approaches towards treating patients with ALS and you'll be able to describe these two approaches that will guide assessment and intervention for patients with ALS, and list five common interventions and approaches to treatment used interchangeably by physical, occupational, and speech therapists.

Okay, so amyotrophic lateral sclerosis is also referred to sometimes as Lou Gehrig's disease, and that's because of the famous baseball player for the Yankees who came down with the illness in the 1940s. It's fatal, as I mentioned, it's progressive, it's a neurodegenerative disorder and it results in profound muscle weakness and wasting of nearly all voluntary muscles, and including the muscles for respiration. The most common cause of death for these patients is inability to adequately take in oxygen. And when we say amyotrophic, that is referring to muscle atrophy, which is a lower motor neuron sign of the disease, and then lateral sclerosis, that's referring to the degeneration and eventual death of motor neurons and the motor cortex, brain stem, anterior horn cells, and the pyramidal tracts of the spinal cord and this is the upper motor neuron signs of the disease. It's a pretty rare disorder, it only occurs in one to two per 100,000 persons worldwide. It is more common in men until the age of 65, and it does peak between 40 and 70 years of age, although I will say I have actually treated

some patients who have had ALS in their 20s. So although it commonly occurs between 40 and 70 years, I've seen patients in their 20s and 30s as well. There are 5,600 patients diagnosed yearly with the disease in the US, and there is an element to the disease that is familial. We'll go over some of the theories and causes of the disease. Although most of it does relate to genetics, there is a form of ALS that is, when we say familial, meaning like it's definitely within the family lines and is passed down from parents to siblings and so on.

The median survival rate is 32 months from onset of the signs and about 19 months from diagnosis, and we'll go over some of the different types of ALS and the different onsets which, some of them, depending on the onset, could have a little bit longer survival rate and some of them have a little bit quicker progression as well. So I mentioned respiratory signs, ALS causes weakness to the diaphragm muscle and other muscles needed for respiration, and as I mentioned before, respiratory failure is the most common cause of death. And all patients who have ALS are eventually faced with the decision as to whether to go on mechanical ventilation or have palliative and hospice care.

So now we're gonna talk a little bit about the pathophysiology of ALS. There have been a number of different theories as to the causes of ALS but most recently, I'm gonna go over a few of some of the genetic factors that they've just recently discovered as playing a major role in ALS and also have connected ALS with some other neurodegenerative disorders including Fronto-Temporal Dementia and Parkinson's disease. So one of the first ones is TDP-43, and TDP-43 stands for Transactive Response DNA Binding Protein. It is actually a normal human cellular protein encoded by the TARDBP gene. This protein plays an important role in normal neuron cellular function by binding both DNA and RNA in neurons. It's normally present within the cell body of neurons, but with ALS, there's an excessive accumulation of this protein and researchers don't really understand if the TDP-43 is sort of like peeking over, I sort of

describe it as like a cancer-like phenomenon where there's just an overproduction of it or something else that is triggering this overproduction of the protein and it's actually thin, so much of it accumulates that it's found in the intracellular portion of the motor neuron tracts in patients with ALS and in the intracellular portion of neurons comprising the fronto-temporal lobe in patients with Fronto-Temporal Dementia, which we will talk about signs and the relation of that to ALS in a few slides. Another genetic cause of the disease is C9ORF72, and this is a gene that is located on chromosome 9. And this gene is associated also with protein binding for axonal and dendrite growth in neurons and it's normally found in the cytoplasm and the presynaptic terminals of all neurons. Mutations to the C9ORF72 expansion have been linked to a diagnosis of ALS, FTD, and both ALS and Fronto-Temporal Dementia, and when we have FTD here, we're referring to Fronto-Temporal Dementia.

And the patients who have this C9ORF72 mutation usually have a lower age of onset for ALS, they have more signs of cognitive impairment and behavioral impairment, and have abnormal changes in the gray matter of the frontal cortex, and it usually progresses quicker and there's a reduced time of survival. Molecular and genetic link between ALS and FTD have some researchers to believe that the two diseases are a continuum of the same disease. So when you're looking at research for ALS, you'll see some studies who refer to the patients as either having ALS, Fronto-Temporal Dementia, or both ALS and Fronto-Temporal Dementia, but the diseases are so linked, including the cognitive deficits that come with FTD, that many researchers, including Zago, believe that they're actually the exact same disease. I just wanted to point that out. So on this diagram here, I'm gonna use the pointer if I can, you could see the red represents the accumulation of TDP-43, and you see three different central nervous systems. The one on the left is ALS, the one in the middle is a normal patient, and the one on the right is a patient who is diagnosed with FTD. And when we look at the accumulation of where the TDP-43 protein is abundant, you could see, in the patient with ALS, it's in the premotor cortex, down through the spinal cord, and in the anterior

horn cells of the spinal cord as well. The control group, there's normal TDP-43, it's in the intracellular portion of all of the neurons, it's finding RNA, DNA appropriately, and then in the patient with FTD, you could see the accumulation in the frontal lobe, which these patients are going to have deficits, or in cognitive impairment, executive function, things like that, and the ALS patient will have more sensory motor impairments.

Another theory on the cause of ALS is the SOD1 gene, and SOD1 stands for Superoxide Dismutase 1. The gene encodes the enzyme, Superoxide Dismutase 1, and it's present on chromosome 21. The enzyme is actually, it's an antioxidant that regulates superoxide.

And superoxide is a byproduct of oxygen metabolism, that if left unregulated, it causes cell damage. And mutations to the SOD1 gene have been linked with ALS. It actually plays, the SOD1 gene plays a big role in the familial form of ALS that I spoke of earlier where there's a clear delineation from ALS being passed down from parent to their offspring. There are other multiple theories on the causes of sporadic ALS. Sporadic ALS just means that it's just naturally occurring, patients may or may not have the genetic markers for the disease but end up contracting ALS at some point in their lives sporadically. And some of those theories include excessive glutamate within the central nervous system, and glutamate is a neurotransmitter. Other theories point to some sort of neuro-inflammatory process.

There's a lot of thought that stress and being associated with a lot of stressful environments throughout your life can cause a neuro-inflammatory process which could contribute to the development of ALS or ALS-type diseases. Environmental factors can play a role, being exposed to toxins, some on heavy metals as well, meaning like ingesting or having lead or excessive iron in your system can lead to neuron damage and development of ALS. Head trauma, we all know that that has played a big role, we've heard about that, especially with patients with ALS who were former football players, NFL players, have been known to come down with ALS.

Although I will say, although I do agree that when you look at the NFL as a whole and players who have spent a lot of time in the NFL and have had multiple concussions and head injuries, there is a higher proportion of them who do develop diseases like Parkinson's, ALS, and CTE. I personally have not, even at eight years at the ALS clinic, have treated a patient who we thought developed the disease because of playing football or that they played collegiate or NFL football. And even when I look at some of those studies, especially with CTE, which is Chronic Traumatic Encephalopathy, which is a type of brain damage from repeated concussions and head injuries, that disease is actually also associated with the TDP-43 displacement that I spoke of earlier, and even in advance forms of CTE, it may actually mimic ALS, both in terms of the cognitive behavioral deficits and some of the neuro motor impairments that these patients have, and this is caused by brain atrophy from repeated concussions and head injuries. When I look at some of these studies, the point I was making, when they look over brain samples from these former NFL players who have had CTE, I mean, I immediately think that, well these brains are being donated to science to be researched, and yes, all of them had signs and symptoms of CTE, and therefore, they most likely did have the disease, but what about the players who may or may not have had signs of CTE, would do their brains look like from a control standpoint?

Did they had normal functioning throughout their entire lives or did they have some minor forms of cognitive or neurological impairments, and what do their brain matter look like upon death? Did they have that same atrophy that players who obviously had CTE and signs of the disease? Some other factors that have been thought of as causes of ALS are excessive physical activity. A lot athletes, marathon runners, just athletes in general who are constantly engaged in physical activity, and then, of course, I talked about the genetic predisposition. A lot of the types of patients that I've treated, who had ALS, they've been involved in military services, or a lot of police officers I've treated, who have had ALS, which I think goes back to the neuro-inflammatory process and exposure to a lot of stress. It seems like these former veterans, these are people

who have faced combat and been in war zones and in war situations, and of course, police officers obviously have a very stressful job as well and I think that that is one of the causes, at least in my experience, of treating this population, just having a job that's very, very stressful. And I'm just going to take off my pointer there. So other signs of ALS. We are gonna talk about lower motor neuron involvement, upper motor neuron involvement, bulbar signs of the disease, cognitive impairments, and respiratory involvement.

So when we're talking about upper motor, lower motor neurons, for upper motor neurons, we're talking about neurons in the brain and in the spinal cord and all of the neurons contained in those areas. When we're talking about lower motor neurons, we're talking about the axons and dendrites that exit the spinal cord and innervate the muscles, and there are different signs and symptoms of upper motor neuron and lower motor neuron involvement with ALS, which we are going to review shortly. All right, so lower motor neuron involvement causes asymmetrical muscle weakness and atrophy, and the lower motor neuron signs usually start off as loss of muscle strength in the extensor muscles of the fingers and wrists, the intrinsic muscles of the hand and dorsi-flexors of the ankle. Functional impairments from lower motor neuron signs typically involve tripping during ambulation, difficulty button shirts, difficulty writing and typing, and some difficulty holding and turning a key.

One of the first signs that I typically saw was usually the dorsi-flexor weakness, patients talking about, just random tripping because of gait disturbances from being unable to dorsi-flex their ankles, causing their toes to get caught on the ground or uneven surfaces, causing them to trip. Other lower motor neuron signs are fasciculations. So fasciculations are very small localized and involuntary muscle contractions. We all have fasciculations from time to time, they normally occur in most humans and it's due to a spontaneous depolarization and repolarization of the nerves and it causes localized muscle twitching. And when you're seeing a patient with ALS,

it's different than the normal fasciculations you or I might experience for a short period of time. They're more pathological in nature, you can see them and feel them. When I am thinking back to my days at the ALS clinic, when I'd be testing manual muscle, when I'd be doing manual muscle testing on patients, a lot of times, if I'm looking at the strength of their wrist and looking at wrist extension, I can actually see, and if put my hand on their wrist extensors, it would almost feel like the muscle underneath the skin was crawling because of the very localized twitching that was occurring at the individual muscle fibers, you could actually see, in some cases, some of these muscle fibers just moving and it almost looked like the skin was crawling, like there was something underneath it. Other symptoms are cramping. So, especially when patients overuse weak musculature from ALS, they will experience cramping, which just to intervene in that would just be to just put a passive prolonged stretch on the muscle, usually takes care of the symptoms of cramping. And the other symptom for lower motor neuron disease is hypo-reflexia, so a decreased response to reflexes. So now we're going to move on to upper motor neuron signs of ALS.

The most common upper motor neuron sign is spasticity, and spasticity occurs due to lesions of the upper motor neuron, of the primary motor cortex. The primary motor cortex, as we know, is responsible for all voluntary and purposeful excitation and inhibition of the lower motor neurons. When there's damage to the primary motor cortex and there's degeneration to that area of the brain, there's loss of control of lower motor neurons, which causes them to fire and the nerves depolarize and for muscles to fire and contract spontaneously, which is what causes spasticity. When we're referring to spasticity, this is resistance to passive range of motion that is velocity-dependent. So the quicker you try to move, let's say you're trying to move someone's arm from elbow flexion to elbow extension, you're going to feel a resistance to that motion and that is due to upper motor neuron signs of ALS. We actually, we can grade the degree of spasticity in the upper motor neuron signs that patients with ALS are experiencing by using the modified Ashworth Scale. Now all of us have, there's normal muscle tone

and all of us do have some degree of muscle tone, which is important. You want to have that because our muscles are in a state where at any given point in time we can make them contract for a purposeful movement as we complete tasks and activity. And all of us probably are zero on Modified Ashworth Scale, meaning that there's no pathological increase in our muscle tone. When you move on to a level 1 in the Modified Ashworth Scale, you're gonna feel a slight increase in muscle tone, you're gonna feel like a catch where it feels like there's some resistance, and then you'll feel the muscle sort of relax as you continue to move the muscle through the range of motion towards the end of the range of motion.

Usually, that catch happens more than halfway through the available range of motion in a muscle and you'll feel the release as you continue to move the joint. If you have a 1+ on the Modified Ashworth Scale, you'll feel a slight increase in muscle tone, it'll be the catch, but then you won't feel that immediate release. There'll be minimal resistance throughout the remainder of the range of motion, and again, that catch will usually happen about 50% of the way through the available range of motion of the joint. You'll feel the catch, but then you'll feel resistance throughout the available range of motion of the joint. If someone was a 2 on the Modified Ashworth Scale, you'll feel a more marked increase in muscle tone. It'll be throughout most of the available range of motion but you can still move the joint freely.

You'll feel, throughout the range of motion, there'll be some resistance but the patient will still have full range of motion. But once you get to a 3 on the Modified Ashworth Scale, that's when there's considerable increase in muscle tone, passive movement will be difficult. Patients at the 3 would still have most of their available range of motion, but again, it's very difficult to achieve full range of motion. Patients who are a 4 on the Modified Ashworth Scale, this is where there's significant rigidity, and both in flexion and in extension, and most patients who are at a 4, they would not have full range of motion. You might be only able to get to 50% of the available range of motion,

possibly 75, but there's definitely a loss in range of motion due to increased spasticity. And if you're unable to test, you would code it as a 9 for that joint. Other signs of upper motor neuron disease in ALS, hyperreflexia. So, unlike with lower motor neuron, where there's a hyporeflexia, upper motor neuron signs have hyperreflexia. So patients are more prone to having increased reflexes, who have ALS, in upper motor neuron signs, and one of the most common hyperreflexia signs is the Babinski sign, and that's where you're placing deep pressure on the lower foot, which causes the first toe to extend and abduct. Bulbar signs of ALS. So when we are referring to bulbar signs, we're talking about deterioration of the cranial nerves coming off of the brain stem, and patients who have bulbar signs of ALS have facial weakness, tongue weakness, difficulty swallowing, dysphagia, dysarthria, and sialorrhea, which refers to an excessive production of saliva, and really, an inability to manage their secretions.

In my experience with treating ALS, I remember some patients who had bulbar onset ALS, and in talking with them and listening to them, I really did not detect any dysarthria, and their speech seemed rather normal and these patients had very normal manual muscle testing, very normal strength, their gait and ambulation and transfer abilities seemed normal, they're working full time, able to drive a car, and when I'd asked them, like, what brought you to the neurologist, like why did you think that there was something wrong? And I remember the one patient just, you know, telling me it was they felt like their tongue was swollen a little bit and it just felt slightly difficult to move, especially just rapidly, like they felt like they were constantly testing it, trying to move their tongue side to side and that's what made them think that there was something wrong, and when they came in, saw the neurologist, went under a lot of neuroimaging studies and some other tests, and yeah, they were diagnosed with ALS. Other signs of ALS, there are cognitive signs to the disease, and for a long period of time, really up until about 2004, 2006, when researchers were identified that Fronto-Temporal Dementia really goes along with ALS and a lot of patients who have ALS are also diagnosed as having Fronto-Temporal Dementia. So there was a lot of

research that was done into the cognitive profile of patients with ALS, and when the researchers first started looking at Fronto-Temporal Dementia, and this is a specific type of dementia that's actually different than Alzheimer's disease, although it's often misdiagnosed as Alzheimer's disease despite the clear differences. Patients with Fronto-Temporal Dementia have a lot of behavioral and social impairments. So there's behavioral changes and sort of just a disregard for social norms, and for those of you who work with patients who are brain injured or work with a lot of neurological disorders, you might hear the term, well the patient's very frontal, and what we're essentially saying by that is that the patient's very forward, there's a lack of inhibition, they might be saying inappropriate things that they normally just would not say.

You know, pointing out things about someone's appearance or their actions to someone that they don't even know or just met, that's when you would say that someone is frontal. And patients with ALS often display this type of behavior and it's often a vast change to how they were typically functioning. A lot of patients with ALS were holding down, you know, very successful in their job career, very successful in the community, a lot of friendships, you know, successful fathers, mothers, things like that, but then there's a change in their behavior and then that puts a lot of strain on these social relationships, and actually some of the first signs of ALS are these behavior changes cause financial distress because they could lose their job, or they end up being divorced, or they alienate a lot of family members.

There's a lot of apathy that comes along with it as well. It's not associated with as much memory loss as there is with Alzheimer's disease, and the difference really being, between the two disease, Alzheimer's and Fronto-Temporal Dementia are patients with Alzheimer's disease, they often can maintain social relationships and they start to become somewhat aware of their memory deficits and they'll sort of hide that from other people, but often patients with Alzheimer's disease don't have major behavioral changes, at least in the early stages of Alzheimer's disease, unlike it is with patients

who have FTD. So despite patients having, with ALS, also being diagnosed with FTD, if you just look at ALS and exclude those patients that have the dual diagnosis of ALS and FTD, there has been a lot of research on the cognitive profile of ALS. Obviously, there has to be cognitive symptoms that come with this disease because of the upper motor neuron degeneration. You look right where that's occurring, it's right in the prefrontal cortex and into the frontal lobe. So patients with FTD, the cognitive profile, or I'm sorry, patients with ALS, who do not have FTD, this study by Beeldman, which was a meta analysis of the literature. It looked at the cognitive profile of ALS in the absence of FTD, and they described this cognitive profile as speech fluency, language impairment, social cognitive impairment, impairment in executive function, and verbal memory.

So we're gonna talk some more about cognitive signs of ALS, and just wanna get my notes here on slide. So when we're talking about executive functioning, we are talking about anticipation, planning, execution, and self-monitoring. Anticipation would be setting expectations and understanding the consequences of your behavior. Planning would refer to being able to organize and think proactively as to what the results of your behavior would be. Execution would be completing a task from beginning to end, and self-monitoring would refer to having emotional control and adjusting your behavior during the completion of tasks. Executive functions, they're interlinked. So I don't want you to look at this list of what executive functions are and think that these are specific types of attributes of executive function.

It's a process, it's not really reducible to each one of these individual skills. For example, if we just look at executing a set of behaviors to complete a task, you'll often be self-monitoring and adjusting your behavior based on your own feedback, and while that's going on, you're anticipating the reactions and outcomes of your behavior and the consequences of your behavior, which will inform your planning. We could apply it to, for example, driving. If you're driving, you're constantly monitoring your speed,

looking for obstacles on the road, anticipating what other drivers are doing. You're monitoring your own actions, executing maneuvers on the road, meaning turning, breaking, steering and signaling. So executive functioning, although it has these different skills associated with it, it's more of a process and if one of them breaks down, the whole system is going to break down. I'm gonna move on to talking to about what pseudobulbar symptoms are. This is, pseudobulbar symptoms, it's inappropriate laughter and inappropriate crying and often referred to as emotional incontinence. The inappropriate emotion that patients experience, who have pseudobulbar symptoms, it's involuntary. So it could be involuntary laughing or crying and it's often disproportionate to the event. So something just benign happens in the treatment room or in the patient's home and really a normal person would not have a reaction where they would start crying or laughing over what had just occurred.

Patients with ALS, you might see this. It can last several minutes, and I just wanna point this out because when we're talking about social functioning in these patients, this can cause embarrassment, it can cause social isolation, anxiety, depression. When I talked about those behavioral changes that lead to, on the far end of it, divorce or loss of a job, this is something that, pseudobulbar symptoms would be something that would go along with that. It's caused by degeneration of the frontal lobes in patients with ALS, and in my experience, as I'm seeing this coming up, I usually think, well is this appropriate? Should I address this? Is it something we should explore with the patient? Are they in pain? But in most cases, you could sort of tell, as you get more experience working with these patients, that it's really just pseudobulbar. I tend to just ignore it and move on and I found that just simple direction, just helps, you know, with the symptoms, if you just move on to something else, refocus their attention, that in a lot cases the symptoms subside. All right, so now we're gonna talk some more about signs of ALS in cognition. Patients who test positive for executive function impairment at baseline often have a faster rate of motor function decline and death. So when patients are cognitively impaired, and that's usually the first sign or symptom that they

have of ALS, research has shown that typically, there's a faster progression that occurs with this. Other signs and symptoms of ALS, I had mentioned respiratory. That could be an onset of the disease, patients might have some normal gait and muscle function and even normal cognition but they might have some respiratory symptoms which would include dyspnea with exertion or lying flat, a weak or ineffective cough, increased use of auxiliary musculature. So you might see less diaphragmatic breathing and more use of auxiliary musculature for breathing. Daytime sleepiness, that occurs even throughout the disease process as patients start to have more and more difficulty breathing.

One of the signs of hypoxia would be daytime sleepiness or decreased concentration or headaches, and that's usually because of a lack of oxygen or oxygen starvation. ALS usually does not affect bowel and bladder function, internal organs, sexual function, outside of the physical ability with that. Sensation, so normal sensation, that's usually in the posterior horn of the spinal cord, which is not the same area of the spinal cord that's affected by ALS. And also eye musculature, blinking and moving their eyes is something that's usually not affected, and actually, that's an alternate way of communication.

Speech therapists and occupational therapists will work with patients who can use an eye gaze system because that ends up, in some cases, some very pronounced cases, becoming the main area for our patients to communicate by using eye gaze system. So that was a lot of information to take in and what does a typical person with ALS look like? I just have down here, this is a trick question because there really is no standard presentation. When you think about the multisystem atrophy that comes with this disease and the progression of the disease, no one patient looks alike. And when we talked a lot about person factors associated with ALS, so lower motor neuron symptoms meaning like muscle weakness, muscles atrophy, upper motor neuron symptoms, meaning the degree the spasticity or hyperreflexia that patients would

have, we talked about cognitive symptoms. These are all person-related factors and we're gonna talk a little bit more about how we have to look at the person within their context and how they're functioning because that's going to also affect the degree of impairment that patients with ALS face. So there's this multisystem breakdown of person factors but each context is unique to that individual person, meaning their physical environment, their social environment, and that's gonna impact their ability to function in their normal everyday lives.

Diagnosis of ALS. So there is no one test used, so there's not a blood test, there's no one neuroimaging study that can diagnose ALS. It usually is just looking at the signs that are available, showing some sort of progression of these signs, and being able to demonstrate that these signs could not be attributed to any other phenomenon that is going on. They use neuroimaging, just look at where the signs are occurring, what the progression looks like, and that's typically how ALS is diagnosed. The El Escorial Criteria is a widely accepted method for diagnosing ALS, and it mainly focuses, there's been some feedback towards the El Escorial Criteria because it really just focuses on upper motor neuron and lower motor neuron signs of the disease, really doesn't consider cognition, even though we know that that is definitely a part of the disease progression.

And when we look at, this is the El Escorial Criteria right here in front of us, and it really just leads us through all signs. So what I was saying, the EL Escorial Criteria is basically signs of degeneration spread within or between regions and a lack of electrophysiological or neuroimaging evidence for other disorders which would explain observed degeneration. And then there's all these different areas of ALS going from suspected ALS, possible ALS, probable ALS, and definite ALS, and whether or not you have one of those four criteria, suspected through definite, it really all depends on the degree of upper motor neuron and lower motor neuron signs that you have. For example, if the neurologist thinks that the patient has suspected ALS, the patient would

just have, well wouldn't just, but they would have lower motor neuron signs alone in two or more regions. So this would really be gesticulations, hyporeflexia, muscle weakness, or muscles wasting in two or more areas, that's usually asymmetric. So you might find that the patient has right-sided wrist extension weakness and left lower extremity dorsi-flexion weakness. That would lead to a diagnosis of suspected ALS. Possible ALS would mean that the patient has upper motor neuron and lower motor neuron signs in one region or upper motor neuron signs alone in two or more regions, or upper motor neuron signs alone in two regions. Probable ALS is when there's a combination of upper motor neuron and lower motor neuron signs in two regions and some of upper motor neuron signs must be rostral to or above the lower motor neuron signs, meaning that the patient might not have weakness in one or more of their extremities but they might have an excessive amount of spasticity and slow movement in that joint.

And then definite ALS is when there's vast degrees of upper motor neuron, lower motor neuron signs in three regions, including bulbar, thoracic, cervical, and lumbosacral and that these signs are progressive in nature, that there's other areas that are being affected as well. So that is the El Escorial Criteria, which is one of the methods used to diagnose ALS. They'll also look at EEG studies, MRIs, other neuroimaging to go, and if all that points towards ALS, that's what leads to the diagnosis. This diagram will show some of the way classical ALS presents itself. So there's a, one, two, three, there's four different circles here on your, intersecting circles on this screen. The middle is classic ALS, we'll work from the middle out. And classic ALS means that, as we saw in the previous El Escorial Criteria, that patients have upper motor neuron signs, lower motor neuron signs and bulbar signs of the disease. I would also add in there cognitive dysfunction as well, although it's not represented on this diagram. But the way those aspects of ALS present themselves as have it being multifaceted, really depends on the onset. So if we start at the top of the circle, you'll see the letters PMA there, and PMA stands for progressive motor atrophy. So progressive motor atrophy is actually a

diagnosis that a patient could have, and really it just has to do with lower motor neuron disease or weakness or fasciculations or muscle wasting. This would be a patient who they're thinking might have ALS but they only have lower motor neuron onset and it really hasn't spread to upper motor neuron or bulbar onset of the disease. And patients who have PMA, again, or progressive motor atrophy, actually have a little bit slower progression of the disease but eventually, the PMA will lead into the lower motor onset. So you'll see, they've entered the circle of classic ALS and then the lower motor neuron onset will lead into classic ALS where it will progress to upper motor neuron symptoms and bulbar onset of the disease.

If we work over to the right, you'll see the letters PLS on the outer circle of classic ALS, and PLS stands for primary lateral sclerosis. It's actually a diagnosis that patients could have where the neurologist might think that the patient might have ALS, but right now, they're only showing upper motor neuron aspects of the disease. They've got a lot of spasticity, maybe some slow muscle movements because of the resistance to stretch, but they don't have any lower motor neuron signs. But eventually, that PLS will lead into an upper motor neuron onset of ALS, and then eventually, into classic ALS where they will have lower motor neuron onset, bulbar onset, as well as still having the upper motor neuron portion of the disease.

On the lower left-hand corner, you see the letters PBP, and that stands for progressive bulbar atrophy. And this is cranial nerve degeneration resulting in dysphagia and dysarthria, and again, much like PMA and PLS, this is a patient who will present with only bulbar symptoms, they're going to have dysarthria, dysphagia, facial and muscle weakness, tongue weakness, maybe some difficulty managing their secretions, hypersalivation, but eventually, that will lead to bulbar onset of the disease and then eventually into the middle of the circle which would be classic ALS. Okay, so what medications or treatments are available for patients who have ALS? I'm gonna go over three different types of medication, the first being Rilutek, which is the most long, for a

period of time, this was the only drug on the market that was approved by the FDA for ALS treatment. It actually, in clinical studies, has only shown to increase survival by two to three months. It does manage some of the symptoms of ALS, some patients report that they feel a little bit more energy, there's less fatigue, maybe less muscle weakness but really that's very few and far between, as I have said, it really only increases survival by two to three months. And you actually have to take it close to the diagnosis or close to when the signs of the disease first start to occur. If you start to take it later on in the disease process, you won't have the effects of the two to three months of survival.

And really, what Rilutek is, if you think back to the first few slides when we were talking about the possible causes of ALS, where I mentioned that glutamate as a neurotransmitter was one of the possible signs of the, or one of the possible causes of ALS? Rilutek is a glutamate reuptake drug, so it reduces the amount of glutamate and works off of that theory that glutamate is one of the causes of ALS. So this next drug is made by Neuropore Therapies, NPT520. This is a very new drug which was just given orphan drug designation by the FDA, meaning that they're giving some partial approval for this drug to be used in clinical studies and to be used with patients who have ALS. And it penetrates the blood-brain barrier and it decreases neurotoxic misfolded proteins, that's superoxide dismutase 1.

It really takes on the role of the SOD1 protein in taking up the byproduct from oxygen consumption or oxygen metabolism in neurons. If you think back to the first couple of slides that we were on, we talked about the SOD1 protein and how it's basically an antioxidant and protects neurons against damage from oxygen metabolism. And damage to the SOD1 gene causes a decrease in that protein being created, there's less of the SOD1 antioxidant factor, this drug actually takes over for that and provides some protection to neurons from oxidation, at least it's been shown to do that in the animal model so far. I lied and there's actually four drugs, although the Tiglutik is

basically just Riluzole oral suspension and it's the same exact drug as Rilutek. It's a glutamate reuptake drug and it's the liquid form of Rilutek and it's used because some patients can't take the tablet form of Rilutek because of dysphagia. It's administered with an oral dosing syringe. Another drug is, excuse me if I'm not pronouncing these drugs correctly, Radicava, and this is a drug that just removes free radicals in the body and provides some neuro protection. It's similar to that drug we talked about previously, which works off of the SOD1 drug that I, or SOD1 gene that I spoke of earlier, in protecting neurons against free radicals, and it provides neuro protection in the absence of the superoxide dismutase.

Again, working off of SOD1, it's just another drug that kinda works in the same fashion. So what are the physical and occupational therapy goals for persons with ALS? We want to optimize function and maximize mobility and comfort through exercise modification, which we'll talk about in a little bit, stretching, activity adaptation, equipment prescription, and patient and family education. So exercise. I wanna talk about this study, which was completed by Lunetta in 2016. So the purpose of this study was to evaluate the effects of three different types of strictly-monitored exercise programs versus a usual care program on retention of function in a cohort of ALS patients. This was an individual, single blind, randomized control study. And what they looked at is they took, over the course of six months, they put patients into three different groups of exercise.

There was a group that had active exercise that was performed with a physical therapist, in a clinic, and the patients had active exercise, plus they had some level of cyclogometer activity, which is basically was just 20 minutes of upper and lower motor, or I'm sorry, upper and lower extremity cycle activity in a seated position. That was one group. The next group had active exercise without the cycle activity. And then the last group just got normal care which they considered just having passive exercises, passive range of motion performed within a protocol. And then they compared, all

patients, I wanted to say, all patients who were put in the study had a manual muscle test of three or five, three out of five or higher, meaning that they at least had antigravity strength being in this study. And what they did was they measured their ALS Functional Rating Scale, and the ALS Functional Rating Scale, it's a validated rating instrument, it's usually used, it was used at my clinic that I worked at, it monitors the progression, or disability, in patients with ALS. It evaluates bulbar, motor, and respiratory function. It's basically 12 questions, it's scored on a scale, from zero to four, zero indicating no function and four being fully independent. So they're scored on performing various tasks and it can show progression of the disease and it could also be used as a prognosticator of survival. The highest score you could have, of course, is 48. They also looked at patients forced vital capacity, and vital capacity measures the function of respiratory muscles. A normal human can, what this does is it looks at either, sitting or lying down, your ability to inhale as deeply as you can and exhale as much of the air that you took in as possible, and a normal human can exhale three to five liters of air during this test. Maximum amount of air a person can expel from their lungs after the maximum inhalation, that's what this test does.

And if the functional vital capacity is less than 50% of the predicted results, that's usually when you're looking at some sort of respiratory support for patients with ALS, which we will talk about, and functional vital capacity is used to look at the progression of the disease and also can be used, along with the ALS Functional Rating Scale, as a prognosticator of survival time. They also looked at the quality of life of these patients. They used the McGill Quality of Life Questionnaire, and all participants were measured prior to treatment on all of these tests, then monthly, while receiving the interventions, until the six-month period ended, and then again, six months post-treatment. So the primary findings were that the exercise plus the cycle activity group had significantly higher ALS Functional Rating Scales at the end of the six-month treatment program and at the six-month followup when compared to the usual care group. All participants who underwent the active exercise program had a higher ALS Functional Rating Scale

at the end of the six-month treatment program and at the six-month followup when compared to the patients who had just the usual care group. No significant differences in quality of life were found and no significant differences in survival were found, and there were no significant differences in the functional vital capacity as well. So obviously, with these patients, with this group of patients, they did have some degree of a, they retained some level of function as indicated on the ALS Functional Rating Scale by participating in exercise.

But I just wanted to point out this study, although as therapists, you might be thinking like, yeah, great, we're just going to just give everybody exercise right now and every patient I have with ALS, I have to put them on a very intensive, strictly monitored exercise program and this will improve or maintain or optimize their level of function throughout the course of their disease, but obviously that is not the case 'cause there are many, many different factors to consider with patients who have ALS because of this multisystem, you know, just devastating atrophy that these patient have and there are many things to consider when you're prescribing exercise for patients with ALS because it can be controversial and there can be some ill effects from excessive exercise with this group.

So as I mentioned, it can be controversial with patients with ALS. Some studies have actually also shown, in addition to this one, there's some studies that show that there are benefits but then there are some studies that, just depending on the person, it could have ill effects. It could increase the rate of weakness and progression of lower motor neuron symptoms and the muscle wasting that comes with the disease. It could cause respiratory distress, difficulty breathing, further dyspnea, and just a longer time to recover. You think about the fatigue factor. These patients who have, or have been diagnosed with the disease, are obviously going to have, they're gonna just have decreased energy levels, and if you're using up their energy levels for the purposes of exercising, it could affect their functioning when they're performing normal activities

that they would maybe typically be able to do despite having ALS which could include ascending or descending stairs, doing normal EDL activities like bathing, dressing, and toileting, preparing meals, and it could cause them, you know, there could be ill effects where they possibly fall because of excessive fatigue if they're exercising too much. So I just caution everybody to take into account a lot of different factors when you're prescribing exercise. It's important to know your patient. If they have normal manual muscle testing, meaning like at least above a four, I would say, those muscles might have some normal response to exercise and there could be some protective elements of performing exercise to sort of stop the progression or sort of like slow down the progression of weakness.

But patients who are already having very weak musculature, that they're three out of five on a manual muscle test or possibly lower than that or have excessive spasticity, you could have some ill effects from performing exercise and it might just be something that's really not worth the patient's time because you're excessively fatiguing them and you might wanna just work on quality of life and really just optimizing their functioning activities that are very important to them and really starting to know their patient that you don't have to do exercises, just doing a repetitive motion for three sets of 10, with a certain amount of weight, maybe not being worth your time but let's talk about what you might want to be able to do. Maybe you like to go out into the community and you want to go to a shopping mall, or maybe you want to attend a baseball or a football game, maybe you've got a son or a grandson who plays a sport and you wanna attend their little league games, that is activity in and of itself and could have the same effects as performing exercise, so maybe you might wanna focus your time and attention on being able to safely perform those activities, versus doing exercise because something's got to give. If you're doing exercise for the beginning of your day, you might not have the energy to then perform or participate in those other activities that I just mentioned up to your satisfaction levels. So, and the other thing is, if a person is very, like exercise is very important to them and something they've done

their entire life, just, you know, speaking of myself, I do exercise regularly, I'm sure many of you on the phone right now, who are physical therapists, you know, that you will go for runs, you go to the gym, you lift weights, many of you might've participated in 5K runs. There's a lot of focus in our society now on maintaining physical function and being active and exercising, so if this is really important to your patient who has ALS, I would just suggest that you look at their routine and modify it and just make sure if everything they're doing is safe for them to perform. So for example, if a patient has maybe some lower motor, or I'm sorry, lower extremity weakness and you're worried about them walking distances or possibly even still trying to run on a treadmill, maybe they should focus doing some seated upper body exercises instead because maybe their upper body strength is close to normal and functioning and they would get some of that protective benefits from doing activity and exercise with their upper extremity versus possibly hurting or progressing their weakness with our lower extremities.

Other things to consider, stop exercising if excessive cramping or fasciculations occur. These could be signs of a progression of the diseases or some ill effects from doing exercise. So patients really have to monitor if they're doing exercises, are they getting some of these other symptoms of the disease? They're getting more cramps, the fasciculations are increasing. There's been some research that has shown also that eccentric contractions can be worse than concentric contractions. So if a patient's doing, what I would call, if you're lifting weights and doing like a negative contraction where you're slowly lowering the weights, those type of activities have been shown sometimes to cause damage to muscles, that are affected by ALS. In general, I would avoid high resistance exercise and high repetitions, I would avoid doing exercises, especially resistance exercises to the point of exhaustion or to the point of failure. And just general, just using common sense, just monitoring your fatigue level. If you do overdo it one day and you feel as though it's affecting your ability to participate in other aspects of your life, you might wanna cut back or maybe cut back on, the amount of exercise you're doing in one particular session, or even just cutback on the number of

times you're doing it per week. Instead of doing four times a week, cut down to three or possibly two, and also I would not recommend giving resistive exercise to already very, especially very weakened musculature. As I mentioned, anybody below a four-plus or a four, I would just start to think about what it is you're having the patient do and not have excessive exercise, especially resistive exercises to those weakened muscles. Stretching is really important, maintaining range of motion throughout the progression of the disease is very important and it's important, I think, to be on a stretching program from the start of the disease, and you can make that as part of the exercise program. And you know, as you normally would prescribe, hold that stretch for 20 to 30 seconds, you wanna perform three to five repetitions daily. Involve the caregivers, because it really should, you don't want this to, you to be the factor, you meaning being the physical therapist, as being the sole provider of the stretch or the range of motion. Involving caregivers in this aspect of the patient's care, only just enhances that social connection and I think can have positive attributes as well, and plus, the more times it's done, the more times they're in the presence of caregivers and the stretch is being done, the higher chance of maintaining your range of motion throughout the progression of the disease will occur.

You know, we cannot, when muscles are already weakened because of ALS, this is not gonna be a restorative disease, you're not going to prescribe exercises in the hopes that you're gonna have a goal where a patient's gonna improve their muscle strength, meaning like, let's say their hip extensors from a three-plus out of five to a four-plus out of five in the hopes that they're then going to be completely independent with stair ascension. That's not the way you treat ALS. You can maintain the muscle length but you're not gonna really be able to, at least over the long period of time, maintain muscle strength or improve muscle strength. And you also wanna do stretching for spasticity management. Stretching does help with just reducing some of the symptoms that patients would have with spasticity and some of the deficits that do come with that. I'm just gonna take a break for a second here while I grab a sip of water. Okay, we

are now going to talk about the Ecological Model of Human Performance, and the Ecological Model of Human Performance I think is a really good way to sort of demonstrate how we would approach treating, not only patients with ALS, but patients who have any neurodegenerative disorder, and you could use this in other aspects, you can use this for treating patients who have an orthopedic condition, but really anything. It's based in the occupational therapy literature but the researchers who created this theory really wanted to focus on having a common language that OTs, PTs, speech therapists, nurses, physicians, that they can pull from and sort of speak the same language. It uses a lot of the same elements that you might see in the International Classification of Functioning by the World Health Organization, the ICF Model that I'm sure all of you go back to, and I too which that we use the ICF in a multidisciplinary context because I do think that, along with the Ecological Model of Human Performance, really presents that sort of common framework that we could all work from as a multidisciplinary team and see where each one of our disciplines fits into the model in optimizing, you know, just quality of life for our patients.

So the Ecological Model of Human Performance has a few key constructs here. So there are person factors, and when we're talking about person factors, I sort of alluded to this earlier, we're talking about sensory motor skills, cognitive skills, psychological abilities, personal interests, values and experience that are unique and embedded in the person, within our patient. The Ecological Model of Human Performance talks about tasks, and tasks as, we all know what tasks are, they're an objective set of behaviors needed to engage in to perform or reach a goal. And tasks can be grouped together to form activities. So for example, brushing, the activity of brushing my teeth might involve the task of reaching for the toothpaste, reaching for my toothbrush, opening the toothpaste container, controlling my arm movement and my pincher strength as I delicately push out the toothpaste on to my toothbrush, then I'm gonna grasp my toothbrush, bring it up to my mouth and then coordinate movements so that I am then engaging in the activity of brushing my teeth. There's also contextual factors

that go along with the Ecological Model of Human Performance. The context, I spoke about that before, the context surrounds the person, and context includes temporal factors and those temporal factors could be your age, your lifecycle, health status. And even expectations, your own expectations, expectations of others. There's physical factors that are in the context, and physical factors are different than the person factors because this is the environment, these are the physical elements and objects around us, that are surrounding us in our home environment, in our work environment, in the community in which we live. There are social factors, and these are our relationships and the norms within those relationships. What type of friends do we have? What type of family members do we have? What type of coworker or colleague relationships do we have? What are the norms of those relationships? What are the strengths of those relationships? Are these deep, vetted long-term friendships or are these casual acquaintances?

And then there's also cultural and environmental factors, and this is really just the beliefs and values of the society in which we live. And different environments could have different cultural factors. Living in Philadelphia, there's different environmental and cultural factors, beliefs and values, and I mean, they're probably pretty much the same, but it might be a little bit different in an urban environment than it is in the Midwest, and certainly, there would be different cultural beliefs and factors amongst countries. So I'm just throwing it, like in India, there would be different cultural beliefs and environmental factors than there would be in Pennsylvania, where I'm at right now. And then, the Ecological Model of Human Performance also talks about performance. Obviously, it's part of the title of the theory and performance refers to the person's engagement in tasks within that context. The core assumptions of the EHP. So the core assumptions state, Winnie Dunn and her researchers, who created the Ecological Model of Human Performance, state that it is impossible to examine a person outside of the context in which they live their lives, and I talked about that earlier. We are constantly surrounded by our context, and even if we move across the country or go to a different country, or

we move jobs, we change jobs, we move out of our houses, we're just moving into a different context, the context changes. We can get a new set of friends but it's just a different context and it's always going to be there. Performance occurs as the person engages in a transaction with his or her context to engage in tasks, and then combination of specific tasks, as I talked about earlier in my example with brushing your teeth, make up roles and occupations. Context can enable and promote, as well as obstruct and inhibit an individual's performance and performance range. When you're thinking about a person who has ALS, they may be able to perform a lot of different tasks and function very well in a one floor apartment, that's small, but you place that very same patient within the context of a physical context of an environment that's two or three story home, that has lots of steep stairs and a bed and bath on the second floor, their level of function might look vastly different. Or a person with lower extremity weakness, who has a lot of lower, maybe modern furniture, is gonna have a lot of difficulty doing sit to stand off of that furniture versus higher furniture, which might make the task a little bit easier.

All of us have a performance range within our context, and it really just has to do with how well we are able to interact with our context, that our physical abilities, our person skills, our cognitive skills, our physical skills, and everything that makes our values, our beliefs in ourselves, if we're able to engage in the context around us, engage with the physical environment around us, engage with the social environment around us, the more effective that transaction is and the more effective the transaction between the person factors and the contextual factors, the more extensive the performance range is for the completion of tasks. If a person is trying to engage within a context where either their physical skills are not a match for the context, they're gonna have a lower performance range. For example, if I was going to go out to run a one-mile race against someone who never trained, I might perform well in that task because the context of the activity matches my abilities. But if I was going to go out and run a 10-mile race against a person who runs everyday, competes at an international level, I would not do

well in that context. And then independence occurs when the person's wants and needs are fulfilled. The Ecological Model of Human Performance model views the use of assisted devices or support of others as resources or tools to achieve independence. So within this model, it comes out and states that anything we as therapists do to improve that contextual relationship between the person factors and the context, that transaction, whether it's tools or adapting the manner in which a person is performing a task, a person who uses a power wheelchair, changes their environment from a two-story house to a one-story house, uses assistance of another person to help them perform sit to stand, or any aspect of bathing or toileting or dressing, those are just tools, it's just a resource available to perform the task and as long as your needs are being fulfilled to your satisfaction level, then that transaction is occurring as intended and you're improving the performance range. This is a drawing, it is a model of the Ecological Model of Human Performance, and you could see there are two circles with an arrow there on the left-hand side.

The larger circle is the context that surrounds the person. Those are the temporal factors, the physical environment, the social factors that surround us at any given point in time. And then the smaller circle is the person. That's where all of your skills, your person-related skills, your strength, your visual motor, your coordination, your energy levels, your beliefs, your values, all of those factors that are embedded in the person, are contained within that circle. The arrow represents the transaction that the person is having with the context, and then outside of the context, you see all of these various Ts. And the greater that transaction, that arrow, where the person is interacting with the context, the wider the performance range. You could see there's two lines there and those lines could be very wide and engulfing a lot of different Ts. This diagram seems to show a fairly wide performance range, we're gobbling up a lot of tasks, but if that transaction was not occurring very efficiently, we might be only a few tasks within our performance range. So it really all depends on that context in that transaction, I'm sorry, the transaction between the person and the context. I do wanna point out here

that when you're treating patients with ALS, the goal might not always be to widen that performance range as much as possible because you have to think about the larger context here, and I'm not just talking about, thinking anticipatory, not just the context that's immediately surrounding the person but thinking about what the effects are of performing an activity to the highest range of the person's ability. So for example, on any given normal day, a person may have the goal of, well, I want to be able to complete my ADLs, my morning routine as independently as possible and be able to go downstairs to have breakfast with my wife before she goes to work. However, if later on that day, the expectation is that I'm going to have to travel to a doctor's appointment, let's say I'm going to the ALS clinic that day, so I have this extra activity that's going to be added into my day that's different than any other normal day, or let's say later that day, we're going into fall and I work for the University of Pennsylvania, so let's say that I want to go to see the University of Pennsylvania Quakers take on the Yale Bulldogs, I may want to narrow my performance range for the ADL portion of my day because I don't wanna use up all of my energy, because knowing later on during the day there is going to be expectations placed on me within my context that I'm not used to because I might have to, if I'm capable of getting in and out of the stadium, maybe doing some stairs.

Maybe I'm in a power wheelchair, but nonetheless, I'm going to be seated upright throughout the duration of the football game, and the context changes and in anticipation of those extra expectations being placed on me, I may want to narrow that performance range and accept more help with ADLs and feeding and getting ready to conserve energy so that I could then have a wider performance range later on in the day. And as physical and occupational therapists, we have to know our patients, we have to get to know them well to know what their interests are so that we can help coach and make these recommendations to our patients and family members, as they progress throughout the disease. And in a few slides, I'm gonna talk a little bit more about our relationship with our patients who have ALS because you have to think

about it as a very long-term relationship, you should want to have a therapeutic relationship with your patients that lasts for a long period of time. And I do not recommend treating patients with ALS on a short-term basis where maybe you're just making a few recommendations for some activity modifications or giving them some exercises as appropriate as they may be the time, and then leaving that patient. You should have a feeling of wanting to be in that patient's life to be able to optimize, and I use the term optimize because, again, we may not always want to maximize the performance range but we wanna optimize the performance range for our patients appropriately, and then teach upon them ways that they can anticipate this themselves and then make the appropriate decisions and appropriate approaches to their activities throughout the day so that they're getting the most out of what they want to be able to do and having a higher quality of life as a result of that. Another approach, theoretical approach, to treating patient with ALS includes the neuro-functional approach.

This is a theoretical framework that was originally designed by patients who had traumatic brain injuries and who have cognitive, severe cognitive behavioral and executive function deficits. I think that this is a good fit with the ALS population as they too can have severe motor, cognitive, and behavioral and executive functioning deficits that are not expected to improve or get better. ALS, as we've learned about previously, it's a progressive disease and this is, I'm gonna go through why I think the NFA approach is a good approach. So the theoretical principles of the NFA, which was developed by Giles, is cognition and function are related but the constructs of cognition or function are not reducible to each other, meaning that much like when I talked about the executive functions and I was talking about those individual executive functioning skills, which, you know, we talked about anticipating, you know, we talked about self-monitoring, and executing, and planning. You can't remove cognition from a functional task, it's as embedded in there as much as it is the physical function of being able to perform a task and you can't isolate cognition from the performance of a functional task. One cannot assess functional performance abstracted from the

context. So much like the Ecological Model of Human Performance, you can't look at the manner in which a person is performing a functional task outside of the context. I think it's very important, when you're treating patients with ALS, although I did most of my treatment within the context of a clinic, I think it's very important to get into the patient's home environment, the environment in which they are transacting with on a day to day basis so that you can get to know them better and get to know what it is that they're facing. The neuro-functional approach is very big on whatever intervention you are implementing, that it's being done in real-time and within the real environment, not a simulated environment like in an outpatient clinic, which I worked at, or in a rehab facility. You truly don't know what the patient is facing until you get into the actual context at which it is they're trying to transact with, and it just leads to better-skilled interventions that can have a real lasting and meaningful impact for patients. And the NFA teaches us that skilled performance develops through attentive repetition of activities through neural restructuring and it was designed primarily for patients who are unlikely to spontaneously develop independence.

So a lot of this is used over, a lot of this meaning the NFA approach is used over a longer period of time, it's not something that you can start to implement on just when you're treating patients for the first, you know, a one-time session or a one-time consult. You really have to use a lot of motivational interviewing. Assessments would look like a lot of just unstructured observation. Really, the NFA approach teaches us that we should just be watching families and caregivers as they transact in their natural environment, to perform different tasks, and you're just sort of studying how things are occurring and where items might be breaking down. And then you might wanna do some more structured standardized assessments after you do your non-structured observation, that's where you might wanna kinda fine tune your theories and hypothesis of what's happening and look at manual muscle testing, you're gonna look at your spasticity and look at where they are in the Modified Ashworth Scale, look at coordination, you might wanna do some more standardized cognitive assessments.

There are actually, we don't have time to go through them, I don't think so much on this lecture, but there are some cognitive assessments that are out there that are more geared towards patients with ALS and identifying the types of deficits that they would have. You wanna do some structured standardized assessments in addition to your unstructured observation. And then, after that's done, start to talk to your patient about what their goals are, use a lot of motivational interviewing techniques to try to draw out of them what their intrinsic motivation is for performing certain activities and the manner in which they want to perform their activities. How did they define success? What satisfaction are they obtaining from performing their activities, and then that's where you could start to suggest your interventions to these patients. And you might wanna use phrases like, I noticed that you're having a difficult time standing throughout bathing and I think it's affecting you later on in the day because you're using a lot of energy.

And what if I were to say to you, I have some ideas that you could continue to bathe yourself and have the privacy of being in the shower, and you know, you can perform the task of bathing without necessarily standing and the whole thing might be that you're going to get them a shower chair or something like that, and that's how you could start to introduce these activities. And then the neuro-functional approach wants you to, so this slide, I kinda go through there. I'm just gonna jump ahead to this slide to use techniques like overlearning. So the deliberate practice of a task past the set criterion, so that if you intervene and you're gonna have them use a shower chair, that they try it out, and then they eventually, that's how they always use the shower chair until we might have to change that intervention because it might be more difficult for them to use that exact shower chair. But using a lot of overlearning, errorless learning and performing tasks in a specific fashion so that the neuro channels, the neuroplasticity occurs during that phase of intervention so that that's the way they're constantly performing that activity and you're changing behavior, their habituating your intervention so that they're optimizing their performance during these tasks. That

is the neuro-functional approach, to summarize it. You could also use tasks, or I'm sorry, use techniques like chaining. So learning whole functional tasks by utilizing a step-by-step process, teaching the sequential tasks of a step, step of a task until completion. So they might learn steps one and two, and they master steps one and two, and then you add step three and then they learn tasks or steps one, two, three, and after they master steps one, two, and three, you introduce four, and then they learn steps one, two, three, and four and so on. That would be a chaining technique, and doing it in errorless learning means that you're anticipating, you don't allow them to do trial and error with the NFA approach, this isn't an approach where you're allowing them free range to experiment with the thought that through trial and error, they are going to learn the best method of performing an activity, you use errorless learning techniques, they have to do it in a specific matter and without error. You're anticipating the errors and preventing the errors so that they're performing the task as intended without any mistakes from start to finish. That's what errorless learning is and it's been shown with cognitively impaired patients who have brain injuries that this type of intervention just helps patients learn better and that they can learn safer techniques to perform tasks in a more efficient and effective manner than if they're making mistakes throughout the process.

This is just a drawing and I apologize for the way some of the wording's coming across on the slide, within the colored portion of those arrows, and this is my self-made theory for treating patients who have neuromuscular disease, including ALS. This is the ecological approach to performance for patients with neuromuscular disease, and you could see two white circles there. It builds off of the EHP and NFA approaches that we just went through. You have two white circles there and you see the person and the context, and the greater degree of interlock between those circles is occupational participation, which is, I mean, that's an occupational therapy term but I fully believe that that also applies to physical therapy because physical therapists, too, are trying to improve the manner with which patients are participating in activities of daily living

through improving strength and functional mobility. So I think PTs play a role in improving occupational participation as much as occupational therapists do. So the greater degree of overlap between the person in context, meaning the greater degree that the person in context are a match for one another, the greater degree of participation that we have. And we have ways, as therapists, to either improve that match, meaning the person in context match, or decrease the overlap. We may want to narrow, as I've talked about in my example with the I'm going to the football game, maybe you wanna decrease, in some situations, the level of participation, and maybe in some situations, you want to increase the level of participation for your patient. And it really all depends on the situation but there are five different arrows that I have as tools that we could use as therapists to either increase or decrease the level of participation by making a better match for the person and the context. The first is therapeutic partnership. I talked about that earlier.

You wanna have a long-term partnership with your patient, you have to get to know them, you want to understand what their beliefs and values are, as well as where they are in terms of their physical skills that are available to them, those person factors that make that person unique, and start to understand what the context, the different context that they're trying to transact with on a day to day basis and that comes through therapeutic partnership. This isn't, as a therapist with all of our patients, but especially with patients with ALS, you should not want to leave them, you should have the desire to stay with them throughout the progression of the disease and not something where you think, well I have to lead up to discharge or I want to discharge them. You should try to find ways that you can continue to work with them and treat them, even in the absence of traditional payers paying for your services, and some of that might be through different agencies that I'm gonna talk about in a little bit, alternate ways that you could seek reimbursement through, for payers through, for example, the ALS Association and other community tools or agencies or affiliations that might be available out there. That's the purple arrow that we see there. The blue

arrow is enhancing functional cognitions. So you wanna use those neuro-functional techniques that I talked about after you do your assessments, unstructured assessments, standardized assessments, get to know your patient, use motivational interviewing, and you identify some interventions that are going to work for your patient. You want to use those overlearning techniques, error-free learning techniques, to try to implement your intervention so that it's long-lasting, it's meaningful to the patient, that they are successful with performing activities in the manner with which you are going to have them do so, and a lot of this is your energy conservation techniques, use of appropriate DME, utilization of services available to them, caregiver support, those are a lot of the interventions that you would utilize and using it in a way to enhance functional cognition. You wanna collaborate with caregivers and interprofessional staff. Much like that therapeutic partnership with your patient, you wanna collaborate with the caregivers that are associated with that patient and with other interdisciplinary staff, the occupational therapists that are working with the patient.

The social workers, the nurses, the neurologists, you wanna get to know them, you wanna be part of the multidisciplinary team associated with working with that patient. And the other thing is you also wanna support the relationship the patient has with those other individuals. The better off the patient has a relationship with their caregivers, the better their function will be and the better their quality of life would be. Keeping in mind that a lot of these patients do have social impairments. So there's apathy associated with this disease, there's a disregard for social norms. We talked about the prevalence of divorce and families strain on patients with ALS. What I would do, as an occupational therapist, is I often just educated caregivers on the cognitive and behavioral symptoms that go along with the disease so that they sort of understand that this isn't their loved one. It isn't that they have a disregard for you, there's something pathological going on, within their frontal lobe, that is just causing them to act like this, and bringing attention and awareness to that, at least sort of

calms the caregiver and they have a deeper understanding of the progression of the disease and they're able to cope and deal with that better. But anything you could do to increase the collaboration in the relationship that patients have with their caregivers and the multidisciplinary team, the better off the situation will be for the patient. The green arrow is the natural context, I talked about that during the NFA, just performing tasks within the natural context of the home or wherever else it is, the work environment that patients are transacting with. And then match the context to the person's abilities.

That's what we do as therapists, we're constantly doing that, whether we're treating a patient with ALS or a patient who underwent an elective knee replacement surgery. We are trying to match that patient's context to their abilities. So in the theory with treating patients with ALS, what I always tell occupational and physical therapists is it's really just what you do, it's just what you do as a therapist, but you might have to tone up and really like turn up your theoretical approach in everything that you do to the maximum because it's so multifactorial with these patients that you can't just use a biomechanical approach and you can't just take the normal approach that you would use for a patient who has a shoulder tendinitis or tennis elbow, or they're coming to you because they had a hip replacement. It's not that type of disease, there's just so many other factors to consider and these are factors that you have been well educated on and definitely, you have the ability within you to be able to treat these patients, you just have to go back to that and really think through what your approach to this patient population is going to be and just turn up your skill level a little bit higher.

So I'm gonna go through just some common interventions that you would use as a physical or occupational therapist or speech therapist for patients with ALS and just sort of comment on that. We have about 20 minutes left so I think, I'm feeling pretty good right now, I think we can get through the duration of these slides within the allotted timeframe, without keeping you too far over, and you'll still be able to get the

most out of it. So for mobility, when patients start to have gait disturbance, I usually, we, my team, meaning that my physical therapy partner would usually recommend a single-point cane and not recommend a quad cane only because the weight of a quad cane, quad canes tend to be a little bit heavier, so a single-point cane would be one of the first mobility devices that we would recommend. For patients who have foot drop due to dorsi-flexion weakness, we would recommend an AFO, but usually a carbon fiber AFO. Again, because it's lighter in weight and it doesn't affect hip flexion, as well as the dorsi, it compensates for a lack of dorsi-flexion but we don't want it to affect knee extension, knee flexion and hip flexion when the patient's walking because of the weight. We usually just recommend a regular rolling walker as opposed to a rollator walker. Some patients did do okay with the rollator walker and there are benefits to that because it has the seat, but sometimes patient, and you all know this, with the way the rollator walker can laterally move, it does require a little bit more trunk stabilization and arm stabilization, upper extremity stabilization, and use of the hand brakes and some patients just didn't have that degree of strength to be able to adequately control the rollator.

And in addition, putting on the brakes before you sit down, that was a safety concern, both from a cognitive and a physical standpoint of being able to operate those brakes, and because the rollator, the seat on them sometimes they're somewhat low and narrow, we worry about patient's ability to sit on it long-term and then actually be able to do a sit to stand to get up off of the rollator seat to go back to walking. So in a lot of cases, we would just recommend a regular rolling walker and just decreasing the walking distance and having maybe a companion wheel chair, which leads into my next topic, available to them at any given point in time. The great thing about a companion wheel chair, these are the wheel chairs that don't have the large wheels that you would manually push. It's used for a caregiver to push a patient and they are, we say they're lightweight, their lighter in weight, they probably weigh 30, 35 pounds, and some caregivers might have the ability to fold them up and put 'em into the trunk of a car or

into a SUV or a van for the patient so you could take them with them if they were going out into the community, so that the patient could walk up until the point where they're fatigued and then use the companion wheel chair later when they're more fatigued. Power wheel chairs are very important, it's very important to anticipate the need of a power wheel chair only because, yes, most cases would qualify for it, with most insurances, but it does take a while to get measured for and get the power wheel chair, it could take a three to four month period. We usually recommended power wheel chairs that had tilt in space functions as well as considering the type of controls. So is it a regular joystick where you just think about the one singular post sticking up were you might have to use a tripod grasp or a pincher grasp on it to control. There's other types of joysticks, ones like we call it the goalpost, it looks like football goalpost, and the patient can rest the palm of their hand within that and then control it without having to actually grasp on to it with their digits. And then obviously physician recommendations, seating recommendations, roho, gel cushions, seat mapping at a power wheel chair clinic are very important when you're recommending a power wheel chair.

You know, with any of these devices, whether we're talking about a Hoyer lift, a power wheel chair, companion wheel chair, I always went with the standpoint that these are just tools, much as like we talked about with the Ecological Model of Human Performance, that these are tools to optimize your performance and participation in day to day tasks and it's not an exclusive request. I might say to you you don't always have to use the Hoyer lift, the Hoyer lift, just accept, have it there as a tool that's available to you on days where you might be exhausted and you need the help of, you're completely dependent to get back into bed, at least you have the Hoyer lift available to help you and a caregiver who knows how to use it. But if you feel okay to perform a stand pivot transfer and your caregiver's capable of helping you do that, even with moderate or maximum assistance, you can do that, you can continue to do that but at least you have the Hoyer lift available to you. And same thing with the

companion wheel chair, you don't have to use it exclusively, but it's a tool to enable you to go deeper and further out into the community than you might normally be able to do walking independently or even walking with a modified independent with a walker, whether it's a standard rolling walker or a rollator walker. And then even with the power wheel chair, even if you're not gonna use it long-term, or I'm sorry, for long distances out in the community, the tilt and space aspects of it are really important for position control because it enables the patient to have control over changing their positions. As you are all out there faced with the task of having to listen to me talk about ALS for two hours, we've all, including myself, we've shifted our weight, I've shifted my weight here thousands of times, and at the subconscious level, for my own comfort and so that I'm able to adequately stay engaged in this lecture and talk to you all about this topic, you're all doing the same as you're listening to me, but patients with ALS who have advanced weakness aren't able to change their position to their liking and you could think about the discomfort that would come with that.

So having a power wheel chair where you're able to change the angle of your, let's say your hip extension or your knee extension, or put yourself back into a tilted position, would just do wonders for your quality of life and you're just overall comfort as you're either just resting, sleeping, watching TV, listening to a podcast or anything else you might be doing. Patients with ALS, you know, they still have occupations and activities that they want to be able to do and perform and not being able to position yourself adequately is going to definitely impact their quality of life and level of satisfaction with participating in those tasks. Mobility continued. You know, proper and safe techniques with transfers, caregiver education, family education, knowing when to limit or avoid ambulating is a big thing with treating these patients. Also, use of a Hoyer lift. I talked about that earlier, if you don't know how to use a Hoyer lift, it is very simple, just go on YouTube, you can look at any video on how to use it, they are very simple to be used, you can use them in very small and confined spaces as well. So don't think that, oh, they don't have enough room for it, that is not true. I could tell you, from a first-hand

basis, I have demonstrated the use of Hoyer lift in very small conference rooms with a bunch of different caregivers and family members of patients who have had ALS and been able to demonstrate the use of it in very small confines. Activity adaptation, energy conservation, obviously a big role in patients with ALS. Home modifications, you know, and even when you're thinking about the progression of the disease and where they are in the state of the disease, is it worth it to spend the money to have the bathroom modified or could we do some smaller modifications that might not cost as much money. Don't use up all of your resources if it's unnecessary or all of your resources on something that you might only be able to use for a very short period of time if you have a fast progression of the disease. Same thing with driving. We have had some patients who have been successful with having some driving modifications made to their car, but again, when you think about the cognitive, executive functioning of driving, you don't wanna, I don't encourage like making massive expensive modifications to your car with the hopes that you're gonna be able to drive independently throughout the progression of your disease.

Driving is one of the most hard and difficult conversations I've had with patients who have bad ALS. I've done some reaction, sort of assessments, just really unstructured, unstructured observation of their reaction times and their ability to generate movements for a base of maneuvers. Non-standardized assessments looking at these types of things for driving, making recommendations. Ultimately, in some cases, I've made recommendations that they go and pay to have a driving exam at a driving center by an occupational therapist, or a physical therapist, so that at least if we know at this moment that they've been deemed that they could still operate a motor vehicle but really starting to think about future mobility and transportation, making use of community resources for transportation, family members, and if there's a place you want to or need to be able to go, by all means, we wanna be able to get you there but you have to get there safely and we have to make sure that you're not endangering yourself and everybody else in the process of getting to your destination. So driving is

a big important factor with patients who have ALS. Again, just accepting our help of use of assisted devices, we play an important role of that, and positioning to increase efficiency with task participation. Equipment prescription. ADL equipment, use it only if time and/or energy efficient, don't prescribe equipment if it's not gonna be useful. For example, reachers and button hooks are great but, sock donners are great but sometimes they could be limited in patients with ALS. If they don't have the physical capacity, the grip strength to be able to use them, or the cognitive capacity, don't recommend 'em, don't force the patient to use them if it's not gonna be helpful. Some cases, it has been helpful. I've had a lot of patients who have benefited from these activities, but again, don't over-prescribe. This is just a picture of a button hook. You could see the wide large grasp for the patient. Larger in diameter items are easier to grasp on to and you just put it through the outside of the button hole and grab hold of the button and you can pull it through. That helps with buttoning shirts. Other adaptive strategies are just avoid button shirts or keep them buttoned and just put them on like a regular shirt.

Other strategies for dressing, dress the weak arm first, then thread your stronger arm in, then use your stronger arm to thread your head through the head hole and pull it down over your trunk. Those are just some very common occupational therapy interventions when you have asymmetrical muscle weakness either due to a neuromuscular disease or some other sort of neurological impairment, CVA or something of that nature. These are just looking at grasps. So like with self-feeding, a tripod grasp, that would be like a more fine motor grasp 'cause it's a more advanced grasp. Someone who has normal hand functioning might use a grasp like that on a fork but you could see that there, in the background there, is a wider grasp bendable spoon which might make grasping a utensil a little bit easier. You could see that grasp is a little less advanced but might be more appropriate, a cylindrical grasp for patient with ALS. And even further, just losing a low tech adaptive support, proximal support for self-feeding, abducting a shoulder. Right there she's probably at about like 60 to 75%

of shoulder abduction. Maybe you could bring it up to 90%, which might make it a little more easier to perform self-feeding in a more gravity-eliminated fashion. She's having less distance to move the food from plate to mouth, and that's just a very easy, common adaptation for self-feeding. You might have patients who will continue to work. You might have to look at a ergonomic setup and taking into those things that we've all learned about, ergonomics, 90-90-90 meaning 90 degree of elbow flexion, 90 degree of hip flexion extension and 90 degree of knee flexion extension. So finding the right seating positioning for patients to be supported throughout their trunk and all of our extremities supported for typing. If they can't type using their hands, maybe using voice typing software, an ergonomic keyboard, an ergonomic mouse, onscreen keyboard, eye gaze software for typing and communicating can work. Headsets, much like the one I'm wearing right now for answering the phone and communicating if you have to do a lot of phone communication, so you don't have to hold the phone up to your ear and mouth for speaking. Other equipment prescriptions include walkers. The ones with the seats, I talked about a rollator versus a regular walker. Canes, stick with the single point cane. Transfer boards.

So if you have a patient who has difficulty transferring, typically I did not recommend the transfer board or Beasy Board, just because it takes a lot of energy and time, as you know, to use that piece of equipment. They're doing little mini sit-to-stands and it could be a taxing effort on trunk musculature, elbow extensors, hip extensors, to perform multiple little mini sit-to-stands on a transfer board than to either just do the stand pivot transfer with a moderate amount of assistance, with the help of a caregiver, or use a Hoyer lift for the transfer, if necessary. So I typically did not recommend transfer boards. Pivot discs have been helpful. For those of you who don't know what a pivot disc is, it's basically like a turntable for a human. If the person can perform a sit to stand but can't necessarily perform the steps of turning their bottom to 90 degrees to sit down from one chair to another in a standing position, you could use a pivot disc, it's basically one disc on the bottom and then a disc on top of it that turns, and when

the patient stands up, the caregiver can move the patient at the hips effectively turning their body position to about 90 degrees to then sit in the commode or chair, or shower chair, wherever it is they're transferring to. The only thing is the patient has to have some degree of ability to stand and stabilize their trunk during that turning process. If the patient is very, very weak, it's not gonna work, it could be unsafe in some situations. Gait belts are helpful, depending on your preference. I, personally, don't use gait belts, I think holding patients around their hips, at their center of gravity, I find that I get a more secure grasp on the patient, I have better control over them when I'm transferring them, but some people do prefer gait belt. At the very least, it could be used as a cue as to where to guard a patient when transferring or ambulating with them around the waist center of gravity and not holding on to an extremity, like an arm, because, you know, as we know, if a patient starts to fall, if you're holding on to their arm, you're not going to effectively keep them from falling and you might injure them and yourself in the process as well.

If patients have hand weakness, you could recommend hand splints to be worn at night or for a few hours during the day to sort of prevent deformities and contractures. Ankle orthotics, we talked about carbon fiber ankle foot orthotics. Cervical collars are great, a big thing. I usually recommend, there's one open one that's like a headrest, that rests on the clavicle, around the sternum and clavicle, and then there's a little post that comes up that the patient could rest their chin on if they have neck cervical extension weakness. Miami J collars can be used as well and other soft cervical collars but sometimes I think it might interfere a little bit with their breathing, so you just have to be careful and just sort of judge their comfort with that. I don't know, in the upcoming slides, if I do have a picture of that headrest, but I think if you just google headrest, cervical collar, you'll see what I mean. They're widely available on any DME store. Wheelchairs, you know, I talked about companion wheelchairs, power wheelchairs. Don't use their insurance benefit for scooters, I would not do that because they get one benefit for a mobility device and you don't wanna use that up on

something that might not last without having the positioning that a power wheelchair has, so don't use the insurance benefit on a scooter. Again, power wheelchairs, you're looking at tilt and space, looking at types of controls, making position recommendations with that. Pain. Is ALS a painless disease? In my experience, not necessarily. You might see research that says, there's nothing indicative of ALS that causes pain, but because of position, spasticity, cramping, things like that, it can be painful. You know, we use nonpharmacological interventions as therapists, stretching, range of motion exercises, positioning, modalities, if they're helpful. Medications can help but they have side effects but we don't prescribe them, we just look at what's ordered and we educate patients to take their medications as ordered by the physician and communicate back to the physician if there are any ill side effects of it so that it can be maybe adjusted or a different medication can be found by the prescribing physician. As I said, ALS is not a painless disease, despite what you read in literature, but you just find the cause of the pain and try to treat it as best you can. For dysarthria, have the patient speak slowly, say the most important words first. If they can write, they could try writing it down, you could try using the communication board as a low tech device or more advanced communication system, as a high tech device. Spell things out, answer yes/no questions.

Make sure that the patient has a way to call for help if they cannot speak, like a life alert or something like that. For dysphagia, advise the patient to chew food slowly, avoid talking while chewing, avoid thin liquids, avoid foods with two or more consistencies, try smaller and more frequent meals, and the big thing that comes up is if the patient cannot take in enough nutrition, there should be a conversation with their neurologist and any other provider who is treating them in terms of a peg tube. You know, patients who can't take in adequate nutrition is really going to affect their health and really affect how much they progress, not from neurological standpoint but just the signs of the disease, you want them to have adequate nutrition. So a peg tube, if it's expected that their longevity could be a little bit longer with the peg tube, a lot of

patients do consider that avenue. For orthopnea, keep the head of the bed elevated, and once patients vital capacity gets below about 50%, I mention that they would be looking for some form of assistive breathing apparatus. So you're going to use non-invasive positive pressure ventilation. You wouldn't be prescribing this but you might recommend it or at least be aware of it or making recommendations to the physician, like I think they have some oxygen starvation here, they're sleepy, what's their vital capacity? Oh, it's less than 50%, have we thought about using BiPAP and starting that conversation? Patients might wear BiPAP as a device to help them breathe at nighttime or even for periods during the day and it actually, for patients who do use BiPAP, if it's caught early enough, there has been some evidence to show that it could improve survival teams and slow the rate of progression of the disease by use, and it just makes sense because they're taking in more oxygen. We all die because we end up not being able to breathe, whether we have ALS or we're dying from something else, the number one cause of death, whether it's heart failure or anything else, it's always gonna be that you're not breathing and having assistance with breathing for a disease that affects your breathing muscles would only help I think your survival rate.

So catching that early on and using BiPAP and making sure they're comfortable with using BiPAP could have good effects and take away some of those, like confusion and other signs of oxygen deprivation that patients might have. You wanna avoid patients with respiratory infections just because it's just gonna make breathing more difficult. Secretions, you could use cough-assist devices such as an inextuffulator, suction to manage secretions, there are also medications, like Botox, which could decrease the amount of secretions, which it sounds painful because they're botoxing the salivary glands but it has helped, patients have been very satisfied with the treatment because you could just imagine how devastating that would be just to not be able to manage your own secretions in that fashion. Psychological factors, we talked a lot about the cognitive factors of the disease and this is just a slide that sort of just goes through just being supportive, just making sure that they have access to the right providers, mental

health nurses, talking about cognitive and behavioral deficits with family members, being anticipatory about possible behavior changes that could impact finances and things like that, Utilities and bills. Just wanna throw out there, like very impulsive behavior could be a symptom of the disease. We've had patients who have just sporadically ordered massive amounts of items on eBay and on Amazon, having things delivered to the house, sort of like a pathological obsession with doing online and QVC and other like TV-based ordering of items that they would have never purchased previously and using up financial resources in a manner that just does not go along with the person's personality or even if they're just illogical. So it's just something to just keep in the back of your mind when treating with patients with ALS and being on the outlook for these obsessive behaviors and not only just finances but also, even nutrition, just eating like obsessively. We had one patient who just didn't even like sweet foods but then all of a sudden became obsessed with eating ice cream. Just things like that do come up with psychological and psychosocial factors with the disease. It's not like heavily prevalent but it does occur. So your resources are the Veteran's Administration, Muscular Dystrophy Association, hospice, ALS Association, Medicare, Medicaid, Social Security disability, county, state and local resources available, just be aware of what's available in your area because as I said, the greater access the patients could have to you and to your services over the long-term could just be very beneficial to them.

So don't disregard what you're capable of doing and advocating for your patient and what you're capable of doing as a physical therapist for these patients. Don't think to yourself, oh, there's nothing I could do at this point in time. There always is and you can constantly coach and just be there for your patients to make very meaningful changes to, again, with the word optimize, their participation in day-to-day activities. The ALS Association, a lot of them have van transportation programs, a loaner closer where patients can try and use and even keep various DME, including power wheelchairs for use if they don't or cannot afford or get them on their own. There's

accessibility programs where you could do some home modifications, home care they could set up. I work for a large home health agency, we provide a lot of care for patients who have ALS. The home visits conducted by the ALS Association, or even Penn Medicine at Home, where I work, by nurses, social workers, assisted technology specialists and other volunteers and just other resource groups for patients and caregivers. And I am only five minutes over. I could open it up for questions right now. So we do have one question, we have a question and I apologize if I'm not pronouncing your name correctly, from Sau-cey? And how long do the episode last? And then Sau-cey also asked the same question, how to support patient when caregiver or family is not supportive? So patients with ALS, I think I had this in one of the beginning slides, that they typically live two to three years with the disease, but it could be longer and it could be shorter depending on the type of onset. Patients who just have lower motor neuron onset could live a little bit longer. Actually, some of the more familial forms of the disease, where it's definitely passed down from parent to offspring, do live a little bit longer. I've actually have had patients who have had the disease, very slow onset of lower motor neurons for over the course of 10 years.

But in most cases, it's about 30 months, 32 months that patients do live with the disease. I think that's what your question is in regards to how long does the episode last. And then what to do for patients who do not have support? We face this all the time, not just with a patients with ALS, but there are patients with other diseases who do not have support in their homes to help them. They're getting I think home care services involved, or is an important first step. Home health services such as, like again, where I work, Penn Medicine at Home, but it could be Bayada Home Health, I think they're pretty nationwide. Also a very good home health agency as well. They have social workers who can connect patients with different community resources, both from association standpoint, like the ALS Association, or to either state and local resources. The Veterans Administration, despite, and this used to make me , sorry to get on my soapbox, but so angry when all of that information came out about patients

waiting for services at the VA, and I do agree that that did occur but the VA, I am telling you, at least in my experience with working with patients who have ALS, have been absolutely wonderful in addressing their needs in a very speedy fashion. The VA has paid for ramps, for complete home modifications for these patients who qualify for VA services, so my hats go off for the VA and I really did get angry over the negative press they received during that timeframe, that was so politicized. So I think just really connecting patient, the ALS Association local chapters are everywhere. They do a phenomenal job with connecting patients to any type of resources they may need, even having state and local government resources pay for up to 24 hours of in-home assistance with patients, and I'm not saying it's easy but the social workers who work there are phenomenal at navigating the system and getting patients connected to services that they need. So to answer your question, that's what you would do for patients who don't have services. It looks like we have another question here from Kathy. Did MOT get recommendations for PT with respiratory failure? Question six.

Did not get recommendation for patient with respiratory failure, question six. Go back to my slide here. I think I can. Kathy, are you talking about just in terms of BiPAP utilization? I'm not sure what your question is. Did not get recommendation for respiratory failure, question six, oh. I think on the, I know what you mean. Is this on the, on the outcome question, maybe. Recommendations, all right. So on your test, question six is recommendations for a patient with respiratory onset ALS, gait disturbance, a history of falls, and four minus out of five limb strength that would include. What are your recommendations for those patients? So it's either, based upon this information that you have a patient who has respiratory onset ALS, so they already have difficulty breathing. They already have orthopnea. They already have a gait disturbance so they're gonna have lower extremity weakness. They're already falling. So there's a lot of safety concerns, a lot of cardiovascular concerns, they already have pretty moderate limb strength throughout their arms and legs, their limbs are already affected by the disease. Would your recommendation be an exercise program

designed for the patient to maintain endurance or strength, which is A, B, would you recommend use of DME, environmental and social factor modifications and energy conservation techniques designed to optimize safe participation and functional mobility in ADL tasks that are meaningful or important to this patient, as B, or would you assess options for splinting and bracing to compensate for that leg and limb weakness, implement that exercise program to improve or maintain their activity tolerance or endurance, but being a little bit careful not to over-fatigue weak muscles, so what would you recommend for that patient? A, B, or C? That is the question number six. I hope that that answered that question for you and sort of clarified. Based upon that information, what would be your recommendation as the best course of treatment for that patient, A, B, or C? Do we have any other questions? Leave, all right.

- [Calista] All right. I don't see any other questions, Scott. Is there anything else.

- [Scott] I do not have anything else to add, I do wanna thank Kathleen and Krista for setting this up and I wanna thank all of you who have gotten on and listened to this. I really hope you enjoyed it, and if there are any other questions that do come up, don't hesitate, when you're at the slides, my contact information is on there, don't hesitate to email me. I'd love to continue this relationship, if you have any questions in the future, please reach out.

- [Calista] Wonderful, well thank you, everyone, for attending the course today and hope to see you in the classroom again. Have a great day, everyone, and have a wonderful weekend. Thank you again, Scott.