Amyotrophic Lateral Sclerosis

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Differential Diagnosis

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Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig disease is a rapidly progressive and eventually fatal neurological disease which affects the neurons that are responsible for controlling voluntary muscles. The word "Amyotrophy" means the atrophy of muscle fibers that are denervated as the anterior horn cells degenerate, causing weakness and fasciculation to affected muscles. "Lateral Sclerosis" refers to gliosis that replaces degenerated neurons which cause hardening of the corticospinal tract (Wijesekera & Leigh, 2009). ALS is part of a group of disorders called Motor Neuron Diseases which are all characterized by gradual degeneration and ultimate death of motor neurons.

This disease was first described in a series of lectures by Jean-Martin Charcot in the 1860s and 1870s about the clinical and pathological features of ALS. Although the methodology that was available during that time was primitive, his original description of ALS is considered accurate today. Lord Russel Brain and Sir William Gowers were in the United Kingdom and provided major contributions, but labeled ALS as Motor Neuron Disease because their belief was that all patients had pathology of upper and lower neurons. This disease was really overlooked in the New World until Lou Gehrig, the famous first baseman of the New York Yankees developed ALS. He first started showing signs in 1938 and was observed when he batted forty-five points lower than his usual career average. By April 1939, he removed himself from the lineup due to his inability to play. Lou Gehrig died two years later and his name is used for ALS in the United States. At the time, he received experimental vitamin E injections in the Mayo clinic (Gordon, 2013.) By 1969, new techniques were used to diagnose this disease including the use of EMG. Advancements in discoveries and research for ALS slowed down until the 1980s when there was an explosion (Oliveira &Pereira, 2009.)

According to the National ALS Registry, the prevalence of ALS in the United States is 3.9 cases per 100,000 people. The two age groups with the highest prevalence rate are 60 to 69 year olds and 70 to 79 year olds. ALS is most common in people above the age of fifty and is more common among males than females across all age groups. Additionally, whites have a greater chance of contracting ALS than African Americans. Patients who are diagnosed earlier have a slightly better prognosis. The average survival time post onset of symptoms is an average of three years to five years; however, there are a small portion of those who live passed five (Mehta et al, 2014.) Because of the variability in progression time and overall rapid progression, it is difficult to predict how long a person will survive. However, longer survival time usually correlates with younger age, better motor function, limb onset, stable weight, more efficient breathing capacity and longer time between symptom onset and diagnosis (Gordon, 2013.)

In about ninety to ninety-five percent of people who have ALS, the disease is a sporadic form, occurring at random with no clear risk factors. Individuals with this type of ALS do not have a family history and therefore, other family members are not considered to be at risk for contracting the disease. ALS disease can be inherited in about five to ten percent of all cases. The familial type of ALS results from inheritance; one parent carries the gene responsible for the disease. Mutations can occur in more than a dozen genes. In about one-third of all familial cases and a small percentage of sporadic cases, there is a defect in a gene known as "chromosome 9 opening heading frame 72," or C9orf72. In another twenty percent of all familial cases, the mutation is in a gene that encodes enzyme copper-zinc-superoxide dismutase 1, SOD1. (Amyotrophic Lateral Sclerosis (ALS) Fact Sheet.)

ALS is clinically a heterogeneous disease in which even family members with the same gene have different manifestations of the symptoms. Besides for the variety of progression rates,

the upper and lower motor neurons are affected differently and therefore the onset will occur in different areas of the body. Behavior and cognitive disturbances vary as well (Gordon, 2013.)

One mutation can have different symptomology and ALS can come about from many different mutations. This suggests that the disease has many different causes with similar pathophysiologic pathways. The gene mutations all cause motor neuron death through different pathways. For example, an SOD1 mutation leads to oxidative stress, and TARDBP, FUS and C9orf72 mutations cause issues in the RNA machinery. The SOD1 gene on chromosome 21 triggers disease in motor neurons, however, the astrocytes and microglia encourage progression possibly through mishandling glutamate. TARDBP mutations may lead to defects in RNA processing. Mutations in the FUS gene cause patients with ALS to have onset younger than forty years old, usually in the arm, and survive less than two years (Wijesekera & Leigh, 2009.)

While the exact molecular pathway that causes the motor neuron degeneration in patients with ALS is unclear, there are similar pathogenic cellular mechanisms that contribute to cell death after disease onset. The C9orf72 gene is thought to be the cause of the genetic form of ALS. However, in sporadic ALS patients, mitochondria abnormalities in morphology and biochemistry have been reported. The mitochondria of ALS patients show increased calcium levels, implying that there is defective energy metabolism, which could have a role in motor neuron damage (Wijesekera & Leigh, 2009.) Furthermore, there is considerable evidence that inflammatory processes and non-neuronal cells may play a part in pathogenesis of ALS. The final cell death of motor neurons in patients with ALS resemble apoptosis. There are also biochemical markers that are found in the later stages of ALS (Wijesekera & Leigh, 2009.)

Although the exact causes of ALS are unknown, there are some genetic risk factors that have been analyzed. Most authors believe that there is a genetic-environmental connection that

cause the motor neuron degeneration. Risk factors have been studied in case control studies which showed that smoking is the only risk factor likely to be associated in the development of this disease. (Wijesekera & Leigh, 2009.) Advancing age is also associated with sporadic ALS. Other correlations include athleticism, specifically professional sports, pesticide exposure and service in the first Gulf War. Additionally, trauma can contribute to all neurological diseases. (Gordon, 2013.)

ALS is a progressive degeneration of upper and lower motor neurons in the brain and spinal cord. It usually begins in the limb or bulbar muscles and spreads until eventually the respiratory myotomes are affected. The motor neurons that innervate the voluntary muscles degenerate and die which include LMN in the medulla, anterior horn of the spinal cord and UMN in the cerebral cortex. Bowel, bladder, and eye muscles are usually spared until the final stages. Muscles become weak and atrophied due to the degeneration, leading to a person's death. Death from ALS is most commonly caused by respiratory failure. Some early signs of respiratory failure include morning headaches, orthopnea, weak cough, and exertional dyspnea. As the symptoms of ALS progress, the patient will exhibit shortness of breath even during the most simple tasks like dressing and eating, and eventually even during rest. (Gordon, 2013.)

Upon attack of the lower motor neurons, symptoms include fasciculation, cramps, muscle atrophy, marked weakness, hypotonia and hyporeflexia. These symptoms are much more disabling than symptoms that are caused by UMN disease which include spasticity, hyperreflexia and moderate weakness. Positive Babinski, clonus, and Hoffman signs as well as emotional changes are also indicative of UMN degeneration (Gordon,2013.)

ALS begins in the limbs; and in two-thirds of the patients, the arms are affected first. When the symptoms begin, they are generally unilateral and focal. Some early signs

include decreased hand dexterity, foot drop, and difficulty walking and lifting arms due to weakness. As muscle function deteriorates the patients may begin to fall and lose their ability to walk. As the disease progresses, and their limbs deteriorate, the patients become more dependent on their caregivers. They also suffer from severe pain due to sensory neuron involvement and contractures from immobility. Additionally, patients' suffer from painful bedsores due to their immobility (Gordon, 2013.)

Patients with Bulbar-onset ALS have a worse prognosis. This type of ALS is more common in older women. The first sign is usually dysarthria followed by dysphagia which can progress to sialorrhea, malnutrition, and anarthria. Patients with this type of ALS usually have an atrophied tongue, and they can have axial weakness which causes dropped head, kyphosis, and ultimately poor balance (Gordon, 2013.)

Cognitive impairment in ALS can be described as overt fronto-temporal dementia which occurs in fifteen percent of patients with ALS and in fifty percent if measured by neuropsychological tests. Subtypes of fronto-temporal dementia include progressive aphasia, semantic dementia and disturbances of executive function, language, judgement, personality and behavior (Gordon,2013.) A study in 2005, compared 20 non demented patients with ALS to 18 controls who were interviewed at two different times over a 6-month interval. A battery of tests were designed to measure executive, memory, language, visual-spacial functions and everyday emotion and behavior. On a simple word retrieval test, the patients with ALS became slower over time, and the caregivers reported increased awareness of cognitive dysfunction in everyday behavior. Patients showed more depressive symptoms on the second interview and also emotional lability at both times (Abrahams, Leigh & Goldstein, 2005.)

During any stage of the disease depression and/or anxiety can increase. Patients with ALS seem to have lower depression rates than expected because they generally approach the disease philosophically. (Gordon,2013) A study performed in 2000, wanted to assess depressive disorders and symptoms in patients with ALS. Measurement tools used in this study included a structured clinical interview for DSM IV, Beck Depression Inventory and others as well. 56 patients with ALS and 31 caregivers were interviewed and data was collected. This study concluded that clinical depression or significant depression is not inevitable or a common outcome of a life threatening illness such as ALS. (Rabkin, Wagner, Del Bene, 2000.) However, patients that do experience depression and anxiety symptoms can impair quality of life through decreased sleep, food intake and feelings of hopelessness. (Gordon, 2013.) Apathy is known to be a manifestation of impaired sleep, anxiety, respiratory weakness, fatigue or medical side effects. It is used as a coping mechanism for people with paralysis or a terminal illness. Apathy occurs in a higher percentage of ALS patients than depression (Silani et al, 2011.)

In order to diagnose ALS, there must be very specific clinical findings in order to exclude "ALS mimic" syndromes which lead to diagnostic error in 5 to 10 percent of cases. The clinical findings must include upper motor and lower motor neurons which cannot be suggestive of any other disease. Therefore, the results alone from an EMG for example are not enough to develop a prognosis (Silani e al, 2011.)

The World Federation of Neurology Research Group on Motor Diseases developed "El Escorial" diagnostic criteria to help diagnose people with ALS for research studies and drug trials. The El Escorial uses the characteristics of the patients and then categorizes them into Definite ALS, Probable ALS and Possible ALS. Definite ALS is UMN and LMN signs in 3 regions. Probable ALS includes UMN and LMN signs in at least 2 regions with some UMN

signs that must be rostral to LMN signs and Possible ALS is LMN and UMN in one region or UMN in two regions with no UMN signs rostral to LMN signs. The Awaji Criteria is used for everyday clinical practice. There are variants of ALS that have differing clinical presentations, prognosis and rate of progression. There is difference of opinion as to whether these syndromes should be classified separately from ALS even though they may have similar molecular pathology. For example, Progressive Muscular Atrophy (PMA) is a LMN syndrome without UMN signs. The "flail arm" and "flail leg" variants which are mostly LMN syndrome have wasting and weakness symmetrically either in upper or lower extremity. Also, Primary Lateral Sclerosis (PLS) is a purely progressive UMN syndrome and there is a debate as to whether this is entirely separate from ALS. Sometimes it is difficult to differentiate between PLS and early stages of ALS because during the early stages, they may only have UMN signs (Silani et al, 2011.)

In order to create a clinical diagnosis for a person with ALS, an expert clinician must perform a precise neurological exam and find UMN and LMN symptoms in the same anatomical segment with asymmetrical localization and requires a neurophysiological confirmation done by an expert colleague. There are many tests that are done to rule in ALS and to exclude the possibility of other diseases and disorders. Electrophysiology studies are used to document LMN symptoms in involved and uninvolved areas and also rule out other diseases. Also, nerve conduction studies (motor and sensory) are necessary for diagnosing ALS and also to exclude any disorders of peripheral nerve, neuromuscular junction and muscle that may mimic the diagnosis of ALS. In ALS patients, the motor conduction velocity and distal motor latency are almost normal. Motor studies are significant in excluding multifocal motor neuropathy through

detecting partial conduction block. Sensory nerve conduction studies can be abnormal with entrapment syndromes and coexisting nerve disease (Silani et al, 2011)

Additionally, concentric needle electromyography shows the presence of LMN dysfunction which provides evidence of ALS diagnosis. These signs should be found in at least two of the four CNS regions, brainstem, cervical, thoracic or lumbosacral spinal cord (anterior horn motor neuron.) Another diagnostic method includes transcranial magnetic stimulation and central motor conduction studies. This is a non-invasive evaluation of corticospinal motor pathways and detects UMN dysfunction in patients who lack these signs (Silani et al, 2011.)

The purpose of neuroimaging is to diagnose ALS and exclude diseases that have treatable structural lesions that can look like ALS. MRI can be significant in revealing lesions in corticospinal tracts in ALS. Furthermore, muscle biopsy is not required for diagnosis of ALS, but are sometimes used to exclude mimic syndrome. Laboratory work-ups are also required when diagnosing a patient with ALS. Abnormal tests include muscle enzymes, serum creatinine, hypochloremia and elevated CSF protein (Silani et al.)

Some diseases can masquerade as ALS and are therefore important to differentiate and rule out. They include: Spondylotic myelopathy/motor radiculopathy, Disimmune LMN syndromes, Hyperparathyroidism, Post-Polio, HIV-1, Paraneoplastic, Encephalomyelitis with anterior horn cell involvement, PLS, Progressive Spinal Muscular Atrophy, Monomelic Sporadic Spinal Muscular Atrophy. There are many other diseases as well. (Silani et al, 2011)

Although there is no cure for ALS, the goal is to increase and continue quality of life and increase survival time as long as possible. Medical management for patients with ALS include medication, multidisciplinary clinics and respiratory support (Gordon, 2013.)

Riluzole is one medication given to patients with ALS and has glutamatergic properties that could reduce excitotoxicity. A double blind placebo controlled study was used to see the effects of different doses of Riluzole on the progression of the disease. The study showed that 100 mg dose was the best benefit to risk ratio and it confirms that it increases the survival of ALS patients (Bensimon et al, 1996)

Additionally, specialty clinics for patients with ALS have been developed in the 1980s and they provide multidisciplinary care. This allows patients to have evaluations by a neurologist every 3 months and for patient and family to meet with expert staff in one sitting. Members of the multidisciplinary team include OT, PT, ALS nurse, dietician, psychologist, social worker and speech therapist (Gordon, 2013.)

Dieticians are important in balancing the patient's nutrition and water intake which become an issue as ALS advances. Due to dysphagia, arm weakness (decreases ability of lifting food to mouth) and hypermetabolism, they develop significant weight loss. Assessing rate of weight loss and calculation of BMI should be used as a predictor for disease progression. Some changes can be made for mild dysphagia which include eating smooth food textures and postural changes. When all interventions fail, and patients cannot increase caloric intake by mouth, a gastronomy can be performed. The gastronomy has benefits including nutrition, improvement of quality of life, and life expectancy, but there is limited information showing the opportune time for the procedure (Gordon, 2013.)

As the disease progresses, respiratory muscles become weaker and they can exhibit symptoms of dyspnea, orthopnea, sleep fragmentation, daytime fatigue and morning headaches. A weak cough caused by diaphragm weakness and bulbar weakness cause poor airway clearance, aspiration and increased secretions. History, physical examination, vital capacity and overnight

pulse oximetry are all used to assess respiratory muscle weakness (Gordon,2013.) The Maximal Inspiratory Pressures, Sniff Nasal Inspiratory Pressure and Transcutaneous CO2 Detector can all detect respiratory muscle weakness as well. There are a few medical interventions to help patients with ALS optimize their breathing. The NIV, Non-invasive Ventilation is used to reduce work of breathing, help with gas exchange, improve sleep quality and enhance survival. Oxygen is usually used together with NIV. The BIPAP can also be used to facilitate physiological breathing (Gordon, 2013.)

Brain Computer Interface Technology is used as a tool to augment plasticity and outcomes for neurological rehab. This technological system is a rehab device that improves the quality of life of patients who have neurological conditions such as stroke, spinal cord injury, ALS and others. As ALS progresses, they can lose communication ability, limb function and eventually eye function and become locked in. This can help them communicate and learn through training induced spasticity to control a computer cursor which will decrease disability. The BCI is still being updated and tested to be certain that it is safe, cosmetically acceptable and accurate. This can also raise ethical issues as to whether the patient whose locked-in should make decisions for ventilator support and other similar decisions. Clinical trials are being performed to determine if the benefits outweigh the harm (Dobkin,2007.)

There are many other procedures that are undergoing clinical trials as well. Antagonism of Nogo is a protein that prevents neurite outgrowth which may improve re-innervation in ALS. Other studies show that exercise programs can have psychological and physiological benefits when performed before muscle atrophy. Additionally, stem cells are being tested as a method of replacing the degenerating cells. Sources of stem cells for ALS include neural stem cells, bone

marrow, mesenchymal stem cells, astrocyte precursor cells, and induced pluripotent cells (Gordon, 2013)

Sinaki and Mulder devised a way to breakdown the disease progression into six stages. This can help guide medical practitioners and physical therapists with the chronology of ALS progression. Stage 1 is the early stage of the disease where a specific group of muscles have mild weakness which can cause limits in performance and/or endurance. Patient is independent in ADL and mobility. PT implications for stage 1 include educating the patient and family about energy conservation and making modifications in the home. The patient should continue his/her day to day physical activities. Exercises to be performed include aerobic activities, stretching affected muscles, AROM exercises and low to moderate weights for resistive strengthening (Bello-Haas, Kloos & Mitsumoto, 1998.)

During stage 2, patient will usually progress to moderate weakness in muscle groups. Foot drop is very common in either unilateral or bilateral lower extremity and may decrease hand dexterity due to intrinsic muscle weakness. The physical therapist should suggest any necessary assistive devices that can be helpful. AAROM and PROM should be done to prevent contractures in affected joints. Patient should also continue AROM, aerobic exercises and stretching as before and strengthening should be continued for unaffected muscles (Bello-Haas, Kloos & Mitsumoto, 1998.)

In the past, Sinaki would advise against any vigorous exercise for ALS patients; however more recently studies have shown that exercise is beneficial for specific muscle strengthening and endurance programs. The physical therapist should prevent overuse fatigue and disuse atrophy when writing an exercise program. Studies have also shown that very repetitive and intense resistive exercises can cause permanent loss of force in muscles that are weak.

Therefore, there must be a good balance for a strengthening program (Bello-Haas, Kloos & Mitsumoto, 1998.)

Four areas of exercise that are beneficial to patients with neuromuscular patients include aerobic exercises, strength, flexibility and balance. Flexibility exercises are important in preventing contractures in patients with these diseases which become painful and extremely prevalent due to excessive time spent in a wheelchair. The benefits of aerobic exercise with a cardiovascular response is similar to healthy patients. In one study, 35 ALS patients were divided into two groups, either using a prolong exercise ergometer or to a progressive exercise test. This study showed that there was an increase in oxygen consumption parallel to heart rate. Additionally, another study with 25 patients were randomly selected to a control group who did any physical exercise and an intervention group who performed aerobic activity including swimming, walking or bicycling. The results showed a moderate exercise program will slow deterioration in patients with ALS (Almeida, Silvestre, Pinto & Carvalho, 20)

A patient with Stage 3 ALS presents with severe weakness which can result in a significant weak hand and severe foot drop. Patient is still ambulatory, but may need assistance standing up from a chair. Ways to keep this patient physically independent and reduce energy expenditure is to use adaptive equipment such as AFO, splints and electrically powered height-adjustable chair. A patient who has a difficult time keeping his/her head upright can benefit from a soft collar. A physical therapist can also advise a patient to use a wheelchair for long distances to prevent fatigue (Bello-Haas, Kloos & Mitsumoto, 1998.)

Stage 4 presents with minimal involvement of the upper extremity and severe weakness of the legs. In this stage, the patient will use a wheelchair and can likely continue ADL. PT implications include, AROM and strengthening exercises to unaffected muscles in order to keep

them strong. AAROM and PROM should be performed in order to prevent contractures. The PT should educate the patient and family about special beds and proper positioning in order to prevent bed sores as mobility decreases (Bello-Haas, Kloos & Mitsumoto, 1998.)

During Stage 5 of ALS, the patient uses a wheelchair due to progressive weakness and decreased mobility and endurance. Upper extremity muscles are moderate to severe and therefore transferring the patient is difficult and may need a Hoyer lift. The caregiver should reposition the patient often and be attentive to skin care in order to prevent bedsores. Pain occurs with immobilized joints and is necessary to address in the treatment plan. Pain is addressed based on the cause. Pain caused by contractures can be helped with thermal modalities, stretching, splinting and soft tissue mobilization. For spasticity and muscle cramping, stretching and massage can help improve symptoms. Pain due to hypomobility can be addressed with joint mobilizations, electric stimulation and thermal modalities. If a patient is having a difficult time keeping their head upright, a semi-rigid collar such as the Philadelphia collar or Newport collar can be worn. A patient with a tracheostomy needs space at the anterior neck therefore a Miami-J collar can be used. Upright head position is important for breathing and to allow eating (Bello-Haas, Kloos & Mitsumoto, 1998.)

A person with Stage 6 ALS is maximally dependent, remains in bed and requires max assist with ADL. Physical therapists can help with proper positioning of the body and educating the family as to proper positioning and prevention of bed sores. It is important that the patient's pain is addressed. During this stage, there is progressive respiratory dysfunction; therefore, a suction should be accessible for secretions. A physical therapist can perform cardiopulmonary techniques using postural drainage, and either self-assisted or manually assisted coughing techniques to promote a cough and mobility of secretions. Airway clearance techniques like

vibrations and percussions can be used as well to mobilize secretions. The goal of this stage is to make them as comfortable as possible and improve quality of life (Bello-Haas, Kloos & Mitsumoto, 1998.)

Patients in the earlier stages that are encouraged to perform exercises, should be cautious not to induce fatigue. They are also advised not to perform continuous long exercise, but rather several exercise sessions for short periods of time throughout the day. Patients should exercise for 30 to 45 minutes a day with ample rest time in between exercise sessions. Exercise can include active exercises, aerobic conditioning and resistive exercises (Bello-Haas, Kloos & Mitsumoto, 1998.)

There are multiple well-known formal assessments used to determine the stage and progression for ALS. The Amyotrophic Lateral Sclerosis Functional Rating Scale assesses a patient's functional status and changes. The caregiver or the patient is asked to rate 10 specific functions on a scale ranging from 0, unable to do the task, and 4, normal function. The ALS correlates positively with upper and lower extremity muscle force and has good internal consistency test-retest reliability (Bello-Haas, Kloos & Mitsumoto, 1998.)

Another scale, the Schwab and England Rating Scale asks the patient or caregiver to measure ADL ability from 0 to 100% (normal). This scale was found to have excellent test-retest reliability and correlate well with qualitative and quantitative changes in patient's functional abilities. In order to assess motor function, quantitative muscle testing using maximal voluntary isometric contractions with a strain gauge tensiometer system. The "PaTa" test, tests bulbar functions in which the patient repeats these syllables over and over in five seconds. Timed 4.6 meter walking test is used to assess lower-extremity weakness and motor control in ambulation. The Modified Ashworth Scale measures spasticity in various muscles which range

from 0, no muscle tone to 4, rigid. Also, the Purdue Pegboard Test assesses pre-hension dexterity and gross motor movements of arms, hand and fingers. Because many patients have respiratory dysfunction, it's important to measure their forced vital capacity, and maximum inspiratory pressure which are evaluated with a desktop spirometer (Bello-Haas, Kloos & Mitsumoto, 1998.)

Case Study:

Bruce is a 45-year-old male married to Tracy and has two boys, one age 9 and the other age 6. Bruce works as an accountant in an accounting firm in New York City. He commutes everyday by train from his home in Westchester, New York. He lives with his family in a beautiful single family home with a nice, large backyard. He is a warm and loving father and loves spending time with his boys on weekends with outdoor activities such as playing ball. He is in good physical shape, goes to the New York Sports Club, three times a week and he enjoys biking for miles at a time. He has a history of carpal tunnel syndrome in his dominant right hand. He also had a right proximal humeral fracture four years ago after falling off his bike. About a year ago, he started experiencing cramping in his right hand while at work. He completely ignored it figuring it was related to his carpel tunnel flaring up. As time went on, he noticed that his hand became clumsy at times when holding a pen at work with a regular grip as well as when he played ball with his kids. He continued ignoring these symptoms attributing it to fatigue and lack of sleep. About six months later, Tracy noticed that while he was walking, his foot would slap down every so often. She explained to him that he must go see a doctor because his symptoms are irregular and peculiar and something must be going on. He went to his regular internist who referred him to a neurologist.

The neurologist performed a physical examination and found weakness and atrophy in bilateral upper extremity and positive Hoffman's sign. He also showed signs of clonus in his right leg along with bilateral lower extremity weakness. Based on these findings, the neurologist diagnosed him with definite ALS because he had LMN signs in at least 2 body regions and UMN signs in at least 1 body region. All other disease processes were ruled out. EMG studies were performed and showed fibrillations and fasciculation in all extremities, normal sensory conduction, low compound motor action potentials in various areas in the extremities, changes in motor unit action potentials, and mild to moderate changes in distal leg musculature. Nerve Conduction Studies and MRI were also performed to confirm the diagnosis. Bruce was instructed to take Riluzole and also antioxidative vitamins with the meals. Him and his family were instructed about the diagnosis and were told about the progression of the disease and each of the stages. They were referred to a social worker to help with family changes and how to address the sudden change of events in their lives. They were also referred to a physical therapist to help them delay the progression of the disease and optimize his quality of life.

Upon meeting his physical therapist, he explained that he had a slapping foot while walking and he had some difficulty going up and down stairs. He also said he had a hard time holding eating utensils with a normal grip. ROM was normal except with thumb opposition. MMT examination showed 5/5 for all lower extremity except hip flexor, right is 4+/5 and left is 4/5. Right dorsiflexors is 3-/5 and left is 3+/5. Shoulder muscle strength on the right side is 4-/5 and left is 4/5. Elbow muscles for the right upper extremity is 4+/5 and the left is 4/5. Ashworth Spasticity Scale is a 1 plus for both legs and 1 for both arms. Balance was assessed by timing duration of unilateral stance, right side was 1 second and left side was 2 seconds. Gait was assessed and Bruce had a positive right foot drop. Hand function was moderately impaired as

shown by the Purdue Pegboard Test where only 5 pegs were inserted in 30 seconds on the right hand and 4 pegs with the left. The FVC and MIP indicates normal capacity. The ALSFRS was taken and the PT concluded that his legs are slightly more affected than arms and that the distal muscles are weaker than the proximal muscle groups.

Bruce was determined to be in Stage 2 of ALS because of his mild weakness in several muscle groups. He has mild problems with handwriting, and holding utensils and also has right foot drop which are all indicative of Stage 2 ALS. He also has complete independence in ADL and mobility.

The physical therapist constructed an exercise program to help with his deficits which included AAROM of the hip flexors, dorsiflexors, shoulder and elbow muscles. The patient would also self-stretch the thumb and plantarflexors in order to prevent contractures. Aerobic exercise with a stationary bike should be performed at 70% of max heart rate. He was explained not to overwork himself to the point of fatigue and was also given a Home Exercise Program. Exercise sessions were 15 minutes twice per day and 10 repetitions for each of the AROM exercises.

Adaptive devices are very important in patients in ALS in order to decrease energy expenditure and be able to maximize their ADL. The use of AFOs were encouraged to help with his foot drop. The physical therapist showed him how to do self-massage when he has cramping in one of his muscles. He was also referred to occupational therapy for a right wrist-thumb splint to be worn during activities and a cylindrical foam to help increase his gripping ability. His family was also referred an ALS support group.

As the disease progresses, Bruce will have to make other household accommodations, have medications altered and have his physical therapy interventions updated as needed. The goal of his medical team is to help slow the progression of the disease, minimize his pain and optimize his quality of life.

Home Exercise Program

Stretching: 30 second hold, 2 sets, Twice a day.

1. Calf Stretch with Towel

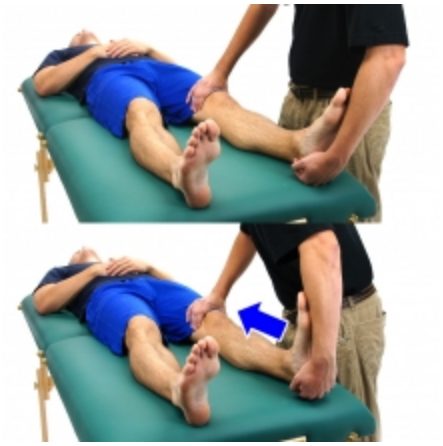
While in a long sitting position, hook a towel under your foot, hold towel with both hands and pull your ankle towards you until a stretch is felt in your calf area. Keep your knee in a straightened position during the stretch.

2. Thumb Extensor Stretch

Grip your thumb with your fingers and then bend your wrist downward as shown for a gentle stretch of the thumb along your forearm.

Strengthening: 2 sets of 10 repetitions, Twice a day.

1. AAROM for Ankle Pump



Caregiver should hold the patient's heel and move the heel up with the toes going up towards the patient and they should both be doing the work of movement together.

2. AAROM with Wand for Shoulder Flexion

Patient lay supine with hands grasping a wand, palms down. Slowly bring both arms with wand overhead.

Precaution: If at any point patient becomes fatigued, exercises should be stopped.

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