CHARLES UNIVERSITY

Faculty Of Physical Education and Sport

Department Of Physiotherapy

Bachelor Thesis

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Case of Study of a patient with Necrotizing Autoimmune Myopathy (NAM)

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Dedication

I dedicate this bachelor thesis to my family in Saudi Arabia my parents, my brothers and sisters who have been supporting me throughout my educational course. It is also dedicated to university staffs that were cooperating with student in a nice way and especially to my professors and teachers for all what they gave me during my study period.

Abstract

Title: Case study of physiotherapy treatment of patient with necrotizing autoimmune myopathy (NAM)

Goals:

To show the full kiensiological assessment, special therapy approaches and therapy effect during 7 sessions. The theoretical part explains the clinical picture, physiological, pathophysiology, etiology, categories, pharmacotherapy and surgical intervention of the myopathy. And the main goal of the practical part is to present the examinations, the therapeutic approaches and the conclusion that were made after the rehabilitation process with the patient.

Methods:

During my clinical practice, we used the main structure for the Kinesiological assessment according to the Prague School approaches. Manual muscle testing according to Kendall (2005) and Janda (2013) are applied as well as joint play examination according to Karel Lewit. We have used active and passive motion, post isometric relaxation according to Karel Lewit, passive stretch and post facilitation stretch according to Janda, also we used mobilization of joint as well as manipulation. Finally we applied final kinesiological assessment to compare the patient situation before and after the therapy and to see the effect of our work.

Results: As a result of our work we found that the therapy course shows few noticeable results in increasing the range of motion in both upper and lower limbs and the function of the both hands and wrists. There was slight improvement in hands, forearm strength that helped the patient while transferring and walking with crutches. However, we could not expect much more improvement even though the patient was excited to go through the exercises but due to her status sometimes we had to stop the therapy session and continued few minutes later or even finish the session earlier than scheduled.

Keywords: necrotizing, myopathy, NAM, ROM, PIR, Physiotherapy, Rehabilitation, and Treatment

Declaration

I declare that I wrote my graduation dissertation (bachelor/graduate) independently, and that I have stated all the information sources and literature I used. Neither this thesis nor any substantial part of it has been submitted for the acquisition of another or the same academic degree.

In Prague, …..... ……

Grandaunt's signature

Records

I consent to the lending of my dissertation for study purposes. By affixing his or her signature the user confirms using this dissertation for study purposes and declares that he or she has listed it among the sources used.

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Acknowledgements

It is an honor for me to have this opportunity to thank the people whom guide me during this work starting by my supervisor Mgr.Michaela Stupková who gave me a lot of her time, which I appreciate it to instruct, guide and advise me to complete this work to be how it is now. Many thanks for Bc. Petr Velišek for his guidance and advises during my clinical practice at RU. I must thank my friends (Maytham, Ahmad, Salman and my brother Mujtaba) who were there all the time for me to achieve this point. My thanks are also delivered to my Saudi Arabian government for giving me a scholarship, which allowed me to study abroad and for their financial support.

Table Of Content

1 Introduction

This work based on a case study of a necrotic myopathy patient. I've chosen this case to be my study case for the bachelor thesis due to my interest in autoimmune diseases and the differentiation of any other skeletal disorders and the lack of physiotherapists who well educated in this field in my country (Saudi Arabia). Therefor, I decided to have this diagnose as my final project. The main parts of my thesis are theoretical and practical part.

The theoretical part consists of a general explanation of the myopathy and its Anatomy, myopathy categories, types and their etiology, Epidemiology, symptoms and the important role of pharmacology intervention, patient's clinical picture and Physiotherapeutic approaches.

The practical part is counted as the main part of my work where it includes a kinesiological assessment (initial and final examination), Therapy progress using some physiotherapeutic approaches which I've learned from the faculty during my study and under the rules and instructions of the institution and my supervisor, Conclusions of both initial and final examinations and the effect of the therapy.

The main goal of the therapy was to strengthening whole body muscles in general and especially neck flexors and hip flexors and extensors for better quality of walking and ADL.

My work was held at the Institution of Rheumatology for two weeks from Monday the 30th of January 2017 to Friday the 10th of February 2017. I had 7 sessions with the patient who had necrotizing myopathy. Bc. Petr Velišek supervised all sessions.

2 General Part

2.1 Myopathy

Myopathies present as pure motor syndromes without any disturbance of sensory or autonomic function. In most myopathies, symptoms tend to be bilateral and affect proximal muscles preferentially. Patients usually complain of difficulty rising from chairs, going up and down stairs, or reaching with their arms. Although most myopathies are symmetric and proximal, there are exceptions to both. For example, inclusion body myositis (IBM) and facioscapulohumeral muscular dystrophy may be very asymmetric. Myotonic dystrophy, distal hereditary myopathy, and IBM may preferentially affect distal more than proximal muscles. In some myopathies, ocular and bulbar muscles may be affected. Deep tendon reflexes are generally preserved or, if reduced, are in proportion to the degree of muscle wasting and weakness. So, we can simplify it by a disease that affect muscles connected to bones (called skeletal muscles), such as the biceps in the upper arm and the quadriceps in the thigh. Myopathies can be caused by inherited genetic defects (e.g., muscular dystrophies), or by endocrine, inflammatory (e.g., polymyositis), and metabolic disorders.

In evaluating a patient with suspected myopathy, it is important to determine whether symptoms are exercise induced. Such symptoms may manifest as fatigability, exercise-induced muscle cramps, or swelling. Patients who present with exercise-induced muscle cramps (see later) may develop frank weakness, swelling, and, if severe enough, myoglobulinuria. These latter symptoms suggest an inherited disorder of muscle energy metabolism. Note that although fatigability is certainly common in myopathies, frank muscle weakness that develops with exercise over a short period of time, if not accompanied by cramps, suggests a disorder of the NMJ rather than a myopathy. Additionally, patients with Lambert–Eaton myasthenic syndrome (LEMS) and some rare patients with myasthenia gravis (MG) present with isolated proximal muscle weakness mimicking a myopathy. In addition, adult onset spinal muscular atrophy, including X-linked bulbospinal muscular atrophy, usually presents with proximal muscle weakness and mimics the typical pattern of a myopathy.(Preston, D. 2013)

Nearly all types of myopathy produce weakening and atrophy of skeletal muscles, especially those muscles closest to the center of the body (called the proximal muscles), such as the thigh and shoulder muscles. Muscles further from the center of the body (called the distal muscles), such as those in the hands and feet, are generally affected less often, so if we would like to be more precisely we can define myopathy that myopathies are a group of disorders characterized by a primary structural or functional impairment of skeletal muscle. They usually affect muscle without involving the nervous system, resulting in muscular weakness. Some myopathies, such as the muscular dystrophies, usually develop at an early age, and others develop later in life. Some conditions worsen over time and do not respond well to treatment and others are treatable and other remains stable. When few treatments are available that address the root cause of the disease, the myopathy is labeled "nonspecific muscle myopathy. (Stanley, J. 2000). Basically, Myopathy came as an explanation of special kind of Myositis, the word myopathy means "disease of muscle." Myopathies are diseases that cause problems with the muscles that control voluntary movements (muscles you can control). These problems range from stiffness (called myotonia) to weakness. They can be mild, moderate or severe. Myositis is the term for any one of a group of inflammatory muscle disorders. These almost always cause weakness and can cause muscle swelling and pain.

2.2 Necrotizing Autoimmune Myopathy

Necrotizing autoimmune myopathy (NAM) is an idiopathic immune-mediated myopathy, along with dermatomyositis (DM), polymyositis (PM) and sporadic inclusion body myositis (sIBM). NAM manifest with subacute proximal-peredominant muscles weakness and elevated serum creatin kinase (CK). Pathologically it is characterized by prominent muscle fiber necrosis and regeneration, but, contrary to the other idiopathic immune-mediated myopathies, it is accompanied with minimal or no inflammation. For this reason, although in the litrature NAM or other immune-mediated myopathies are sometimes referred to as " idiopathic inflammatory myopathies" (IIM), we prefer to avoid the term IIM. (Angelini.2016).

Necrotizing Autoimmune Myopathy is a rare acquired muscle disease believed to

immune based as indicated by Distad, Amato & Weiss (2011). This disease can either be associated with the use of statins or associated with cancer. As opposed to patients with myositis that experience inflammation as well muscle biopsy, patients with NAM do not experience inflammation or may only experience it slightly on muscle biopsy (Understandingmyositis.org 2017).

Although the exact mechanism pertaining to the disorder is not well comprehended, most patients with NAM have autoantibodies directed against 3-hydroxy-3-methylglutayrl coenzyme A reductase (HMGCR) utilized in the biosynthesis of cholesterol (Understandingmyositis.org 2017). HMGCR is an enzyme that is targeted by statins and has been linked to the onset of NAM. However, many patients with the condition have never had statin medications- usually recommended for the treatment of dyslipidemia and risk reduction in cardiovascular disease (Ramanathan *et al.*, 2015). Nonetheless, the availability of antibodies and the response to immunotherapy back the autoimmune nature of NAM as indicated on Understandingmyositis.org (2017).

According to Ramanathan *et al.* (2015), inflammatory myopathies are heterogeneous group of muscle acquired disorders that constitute dermatomyositis, polymyositis, inclusion body myositis, and overlap myositis as indicated by Drouot *et al.* (2014). The recent characterization of NAM involves placing it in a category of severely acquired myopathies, with prominent features of myofibre necrosis that do not involve many inflammatory signs. Since the condition has been lacking outstanding biomarkers, it has over the years been misdiagnosed as a form of myositis with low possible chances of causing inflammation if any as posited by Drouot *et al.* (2014). However, as Distad, Amato &Weiss (2011) expound, studies were done and reports given in terms of descriptions of clinical cases of necrotizing myopathy with microangiopathy and microvascular deposition of complement, the conditions were progressively distinguished from myositis, which is finally classified as NAM (Drouot *et al.*, 2014).

NAM may be linked to a number of physiological components such as anti-signal recognition particle (SRP) autoantibodies aAbs (Drouot *et al.*, 2014). The anti-SRP aAbs are not present in many individuals with since only 4 to 6 % of individuals with acquired inflammatory and/or necrotizing myopathies have it (Drouot *et al.* 2014). Such patients are linked to detrimental clinical types especially those having heart-related diseases (Drouot *et al.*, 2014). In three-thirds of the reported cases of NAM, the conditions have

been linked with the availability of anti-SRP and anti-HMGCR aAbs in the body system of patients. The main role of SRP is to allow the transfer of nascent protein from the cytoplasm to the endoplasmic reticulum for synthesis. However, the availability anti-SRP blocks this mechanism from taking place in the cell (Simons *et al.*, 2016). On the other hand, anti-HMGCR aAbs target the catalytic sites that control the biosynthesis of cholesterol within the enzyme hydroxyl-methylglutaryl- coenzyme A reductase. This affects the functioning of the hypocholesterolemic drugs- statin in the body whereby this enzyme acts as the pharmacological target (Simons *et al.*, 2016).

Although the frequency of occurrences of NAM is difficult to determine, the condition constitutes a whopping 20% of the autoimmune myopathies (AIM) as cited by Simon *et al.* (2016). NAM is a condition that affects adults more than it does children, with women the most affected in the most reported cases (Simon *et al.*, 2016). It is also worth noting that the reactions of affected individuals to this condition varies; there is a group that is presented with corticosteroid resistance (Simon *et al.*, 2016) while in other people the condition may mimic dystrophy leading to a delayed diagnosis as posited by Simon *et al.* (2016). However, the pathophysiology of necrotizing autoimmune myopathy (NAM) is not well discerned due to the absence of animal models to aid in several studies as pointed out by Simons *et al.* (2016).

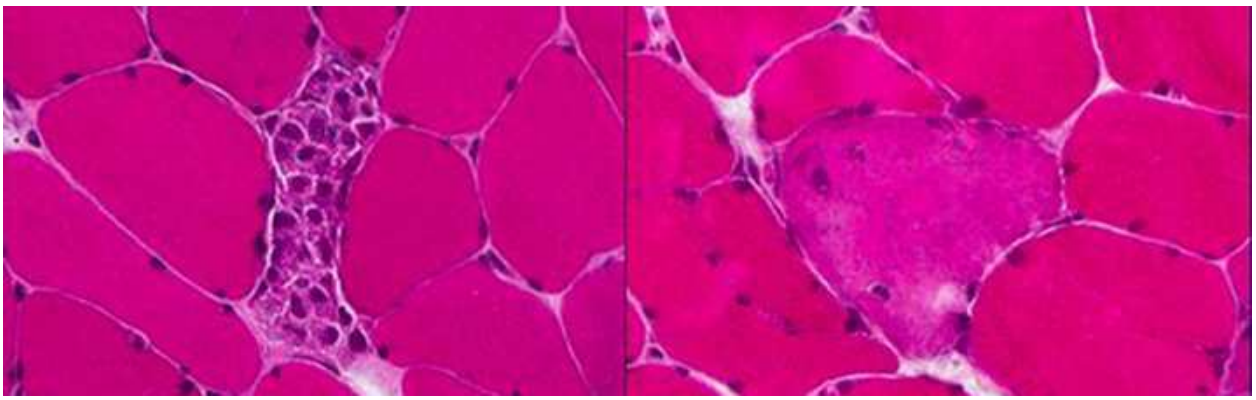

Figure 1: Shows cells split through myopathy

Credit: Advances in Clinical Neuroscience and Rehabilitation (ACNR 2017).

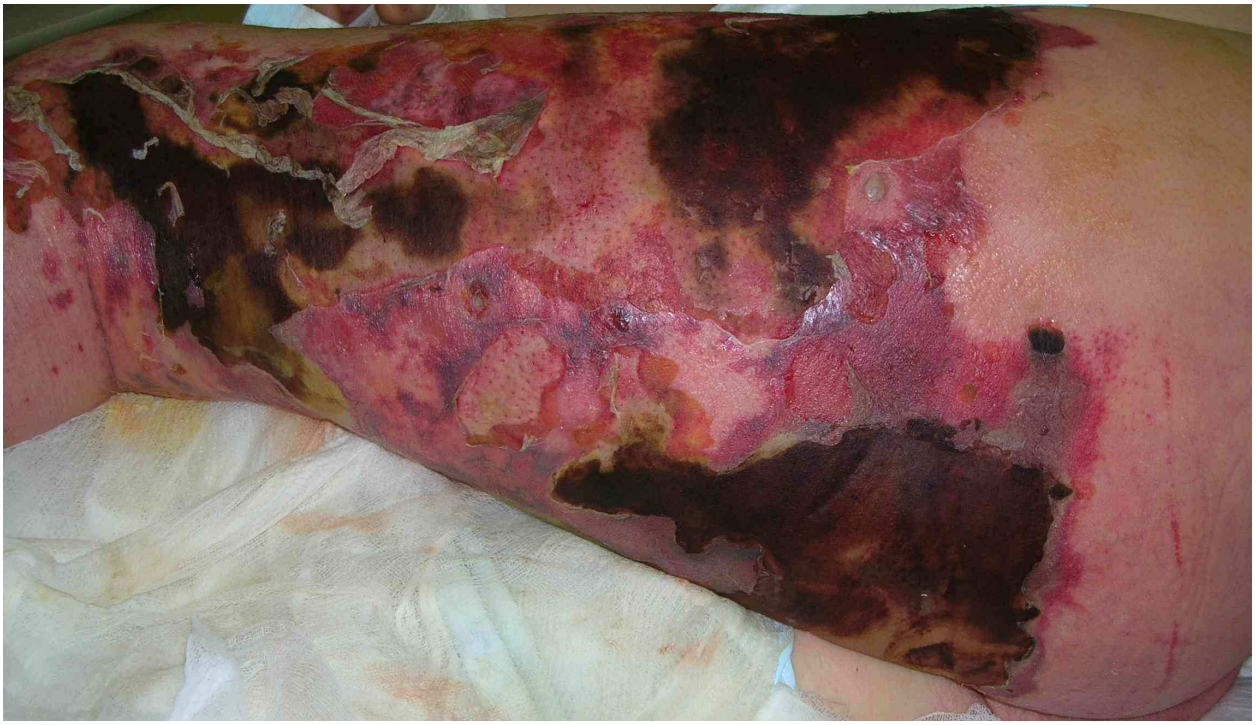

Figure 2: Shows skin of a patient affected by Necrotizing Autoimmune Myopathy (NAM)

2.3 Clinical Features and Epidemiology

NAM is a rare kind of idiopathic inflammatory myopathy identified clinically by acute/subacute proximal muscle weakness, or histopathologically through myocyte necrosis as well as regeneration without notable inflammation (Orpha.net, 2017). The condition though up to date not fully discernable, has several noticeable clinical features as well as epidemiological features that are worth looking into. NAM is highly associated with connective tissue disorders (overlap syndrome) and viral infections (Orpha.net, 2017). Some reports linking it with HIV and Hepatitis C as well as medications – statins in particular and malignancy have also been provided (Nichols *et al.*, 2015).

Most patients with NAM present during adulthood between the second and the sixth decade, but the disease can also manifest in the elderly and childhood. Overall, there seems to be no clear male or female predominance, and different studies have shown conflicting results in this regard. The typical clinical presentation of NAM is of relatively symmetric proximally predominant weakness that progresses sub acutely (over days,

weeks, or a few months) in most cases. NAM often manifests in a more fulminant manner than typically seen in inflammatory myopathies, and more than 50 % of patients develop severe weakness. However, some reported NAM cases have presented insidiously over many months, mimicking muscular dystrophy and making the diagnosis challenging. Lower limb weakness tends to be more severe than upper limb weakness, and patients often complain of difficulties arising from a chair, climbing stairs, or ambulating. Distal weakness is common, although less severe than proximal weakness. We observed distal weakness in 41 % of our patients, mainly affecting wrist and finger extensors and/or foot dorsiflexors. Axial weakness occurs in 30—80 % of patients and neck flexor are more commonly involved than neck extensor muscles. Occasionally, neck muscle weakness can be the predominant clinical feature. Bulbar muscles may be affected with one-third of patients experiencing dysphagia, whereas dysarthria is uncommon (only 6 % in one series). Myalgia occurs in approximately one-third of the patients. Rash is extremely rare in NAM. The characteristic skin manifestations of DM are not observed. However, if a patient has a coexisting connective tissue disease, the dermatological or systemic features of the specific connective tissue disease (e.gi, scleroderma) can be observed. Raynaud phenomenon can occur. Weight loss around the time of presentation of NAM is not uncommon and observed in about 30 % of cases. (Angelini, 2016).

2.3.1 Clinical Description.

The condition has varying ages of onset; with the most age in reported cases ranging from 30 to 70 years (Orpha.net, 2017). The feature that is most striking about NAM is subacute severe symmetrical proximal myopathy, with a markedly elevated Creatine Kinase (Drouot *et al.*, 2014). The manifestations of clinical symptoms are closely related to those seen in polymyositis with severe weaknesses experienced in both the upper and lower limbs leading to difficulties in shifting from sitting positions, climbing stairs, or lifting objects (Orpha.net, 2017). These weaknesses may also be extended to include the neck flexor, pharyngeal, and respiratory muscles as cited on Orpha.net (2017). Other key symptoms also encompass fatigue, weight loss, dyspnea, and dysphagia. Some reported cases related to the condition over the years have been interstitial lung disease

and cardiac involvement (Orpha.net, 2017). NAM's progression is usually severe, but it may be self dependent, and recovery may happen within weeks-months of stopping the causative agent if determined (Orpha.net, 2017).

Statin induced necrotizing autoimmune myopathy (SINAM) can begin initially showing signs that are linked to statin resistance like myalgia or mild weakness, and deadly signs may show up later as the condition progresses within the body of the patient (Babu &Li, 2015). The average time for the symptoms to start showing up is three years; however, the duration ranges from 2 months to 10 years (Babu & Li, 2015). Even though stopping the use of statins as key causative agents have in several studies been shown to indicate improvement following cessation of use, sometimes stopping the causative agent may either provoke the symptoms to show up or lead to a non-substantial reduction in the severity of the symptoms as posited by Babu & Li (2015). Given that several types of statins have been shown to cause SINAM it, therefore, follows that the issue is more about class in statins rather than specific statin effect (Babu & Li, 2015). Contrary to other forms of muscle disease, SINAM rarely occurs in the context of another systemic autoimmune disease (Babu & Li, 2015).

In the majority of NAM incidences, their inhibition of expression of biological components on the surface of major histocompatibility complex-I (MHC- I) hence interfering with some physiological processes within the cells (Nichols *et al.*, 2015). Up regulation of HMGCR in regenerating cells may be the cause of a continued immune activity, even in the absence of statin as posited by Nichols *et al.* (2015). In acute NAM, the onset can take days to weeks as opposed to the statin-induced NAM whereby symptoms can take years to be expressed following either the start or the stop of using statins (Nichols *et al.*, 2015).

Statins may, therefore, trigger myopathy based on immune interference that may persist after drug discontinuation and responds to the therapy that suppresses these immune activities (Nichols *et al.*, 2015). Statins are therefore of clinical relevance in studying the progression of necrotizing autoimmune myopathy (NAM) since they offer the required guidelines in knowing the nature of the condition. Clinical evaluation early enough at the clinical level is important in reducing the morbidity and mortality caused by

NAM as a group of idiopathic inflammatory myopathies (IIM) as cited by Mandel, Malemud & Askari (2017).

2.3.2 Epidemiology

The annual incidences of NAM as well as the prevalence are not well known since the disorder is rare. Up to recently, only about 300 cases have been reported as indicated on Orpha.net (2017). Nonetheless, the current literature indicates that cardiac-based health adversities in patients with myositis vary between 6% and 75% depending on the patient selection modalities, the definition of heart involvement, and methods used to detect cardiac abnormalities (Danieli *et al.*, 2016).

2.4 Etiology and Risk Factors

According to Orpha.net (2017), NAM is highly linked to immunity activities and is activated through the use of statins as therapeutic drugs, connective tissue disease, as well as cancer. Although the actual mechanism is not fully understood, autoantibodies are critical in triggering this condition as well as progressing the disease. Some of the common risk factors associated with NAM include exposure to statins, connective tissue disease (CTD), cancer, and in very rare case, human immunodeficiency virus infection (Kassardjian *et al.*, 2015).

Some of the risk factors that undermine the understanding of this disease are associated with setbacks in muscle histology that lead to poor diagnosis and interpretation of results. This may lead to the failure to distinguish NAM from other myopathies or from inflammatory dystrophies as cited by Dakalas (2011). Some of the errors that can lead to risks in interpretation include the following. For one, there might be a failure to recognize muscle fiber necrosis due to invasion by macrophages as observed in inflammatory dystrophies as well as NAM (Dakalas, 2011). Secondly, there may be a failure to evaluate blood vessel pathology or seek for complement deposits that can be valuable in some cases of NAM (Dakalas, 2015). Nonetheless, there may be a failure to appreciate the

presence of other inflammatory cells that may be of utmost relevance in the diagnosis of certain body disorders as asserted by Dakalas (2015).

Rarely, NAM has been associated with connective tissue diseases, including systemic lupus erythematous, scleroderma, Sjogren syndrome, or an undifferentiated connective tissue disease. In these cases, patients often have multisystemic symptoms or clinical signs suggestive of the underlying connective tissue disease and a compatible serological autoantibody profile. There is no convincing evidence that NAM patients with an underlying connective tissue disease have a different clinical course than other NAM patients, but the small numbers of reported cases preclude reliable conclusions.

Statins appear to be the most common risk factor association in patients with NAM. Several studies have demonstrated an increased risk of myopathy, including NAM, in patients taking statin medications. In patients exposed to statins, who develop myopathic symptoms and elevated serum CK, NAM should be suspected if there is no clinical or serological improvement after discontinuation of the statin. Patients may be on statins for months or years (2 months to 10 years in one study) before developing NAM, and in some cases NAM has occurred many months after statin discontinuation. In these cases, it may be difficult to identify the statin as the underlying trigger.

Interestingly, regardless of disease or drug association (paraneoplastic, connective tissue disease, or statin), there appear to be no significant differences in clinical severity, extra-muscular manifestations, serum CK, or clinical outcome. Statin- associated NAM patients tend to be older and are more likely to have myotonic discharges on EMG, whereas idiopathic cases are more likely to have dysphagia. (Angelini, 2016).

2.5 Types of myopathy

Myopathy is a common term for a muscle disease that is unrelated to any disorder of innervation or neuromuscular junction, with a wide range of possible etiologies. Practically all types of myopathy result in weakening and atrophy of skeletal muscles – particularly those closest to the center of the body (also known as the proximal muscles), such as the thigh and shoulder muscles.

The myopathies can be divided into hereditary and acquired disorders. Hereditary group encompasses muscular dystrophies, congenital myopathies, metabolic myopathies,

mitochondrial myopathies, as well as myotonias and channelopathies. Conversely, inflammatory, endocrine and toxic myopathies belong to the acquired group.

2.5.1 Hereditary Myopathies

Muscular dystrophies represent a heterogeneous group of hereditary illnesses affecting both children and adults, with at least 30 different genes responsible for the disease development. These diseases are characterized by muscle wasting and weakness, with elevated levels of creatine kinase (CK). The diseases show a dystrophic pattern (*i.e.* degenerative pattern with necrosis and extensive fibrosis) and an involvement of the central nervous system.

Congenital myopathies are also a genetically and clinically heterogeneous group of conditions, originally classified according to unique morphological changes observed in the muscle tissue. Still, no necrotic or degenerative changes are present in congenital myopathies (in contrast to muscular dystrophy) and CK levels are often normal. This group of myopathies includes some well-established conditions such as nemaline myopathy, central core disease, X-linked myotubular myopathy and centronuclear myopathy.

Metabolic myopathies comprise a diverse group of disorders, which arise as a result of defects in cellular energy metabolism, including the vital breakdown of fatty acids and carbohydrates to generate adenosine triphosphate (mainly via mitochondrial oxidative phosphorylation). Consequently, the three main categories of metabolic myopathies are fatty acid oxidation defects, glycogen storage diseases, and mitochondrial disorders due to respiratory chain impairment.

Mitochondrial myopathies are also a large group of variegated disorders resulting from primary dysfunction of the mitochondrial respiratory chain and subsequently causing muscle disease. This group of illnesses has a myriad of different phenotypes and genetic etiologies, and can frequently present with multi-system dysfunction. Typical examples of mitochondrial myopathies include severe Pearson syndrome, Kearns–Sayre syndrome and progressive external ophthalmoplegia that can manifest in late adulthood.

Genetic defects in the genes that code for calcium, sodium, potassium and chloride channels in skeletal muscles result in the periodic paralyses, the nondystrophic myotonias, and the ryanodinopathies. Researchers have expanded and clarified the role of gating pore current in channelopathy pathogenesis; hence this group of diseases now includes myotonia congenita, paramyotonia congenita, hyper and hypokalemic periodic paralysis, potassium-aggravated myotonia, as well as Andersen-Tawil syndrome. (Meštrović, 2015).

2.5.2 Dermatomyositis and Overlap Myositis

Dermatomyositis (DM) is prevalent in both adults and young children and highly linked to cancer in adults and calcinosis in children (Nichols *et al.*, 2015). The clinical symptoms of this myopathy include skin manifestations that go beyond muscle weakness. Some skin features for this myopathy include violaceous eruption (Gottron's rash) on the knuckles that may proceed to scaling discoloration (Gottron's papule), purple periorbital heliotrope rash with edema notably on upper eyelids (Distad, Amato & Weiss, 2011); while others are less specific as erythematous rash on the face, knees, elbows, malleoli, neck, anterior chest, back, and shoulders, or periungeal erythema, painful to pressure (Simons *et al.*, 2016).

Muscle weakness invariably involves limb girdle muscles and less frequently respiratory and pharyngeal muscles (Simons *et al.*, 2016). Due to injuries caused to the myofiber in DM, serum is usually found in high levels of creatine kinase (CK). DM is easily distinguished from other forms of myopathies following histological analysis by looking at the levels of inflammation and presence of microfiber necrosis accompanied by regeneration (Dalakas 2011). The pathophysiology of this condition is not well understood given that muscle biopsy shows not only immune cells infiltrates but also C5b-9 deposits (Distad, Amato & Weiss 2011). However, its association with a high number of aAbs is still not clear whether it is such molecules that trigger the release cytokines, take part in endothelial cell adhesion, as well as, migration of B and T lymphocytes responsible for producing Type I interferons (Distad, Amato & Weiss, 2011). These cellular components stimulate the production of pro-inflammatory cytokines and bolster expression of HLA class I and class II molecules (Mandel, Malemud & Askari, 2017).

2.5.3 Polymyositis

This myopathy is generally used to refer to a collective word for idiopathic inflammatory myopathies (IIMs). These myopathies lack specific features that can allow them to be placed into one of the four existing categories or they may be negative for myositis-specific antibodies as cited by Mandel, Malemud & Askari (2017). Patients with this type of myopathy share symptoms similar to those with DM in terms of having subacute proximal symmetric muscle weakness; however, the patients under this category tend to lack the characteristic rashes seen in dermatomyositis (Mandel, Malemud & Askari, 2017). The difference is seen in the instances that involve serum CK elevation up to 50 times higher than the normal in the subacute active phase as cited by Mandel, Malemud & Askari (2017).

2.5.4 Anti-Synthetase Syndrome

This myopathy is divergent from other subcategories of myopathies in that it is associated with positive anti-Jo1 myositis –specific antibodies in approximately 75% of the patients (Understandingmyositis.org, 2017) However, some cases have posited that muscle weakness and biopsy are similar to those seen in dermatomyositis (Mandel, Malemud & Askari, 2017). This condition is highly associated with arthritis, Raynaud's phenomenon, fever, and "mechanic's hand" (Distad, Amato & Weiss, 2011). It presents with it a unique and singular entity on its own as remarked by Mandel, Malemud & Askari (2017).

2.5.5 Sporadic Inclusion-Body Myopathy (sIBM)

This type of myopathy is mostly acquired in patients with above 50 years of age (Simons *et al.*, 2016). The condition has a slower natural course with a progressive muscular weakness deteriorating on a yearly basis leading to ambulation loss, respiratory muscle weakness as well as dysphagia. The condition affects striated muscle with a characteristic pattern affecting quadriceps, fingers, deep flexors, and possibly pharyngeal muscles (Simons *et al.*, 2016).

As posited by Dalakas (2011) pathological criteria have been of major importance to retain the diagnosis of sIBM. The new guidelines no longer need bringing together all the pathological signs (Dakalas, 2015). Inflammatory infiltrates exist with mononuclear cell invasion of non-necrotic muscle fibers (partial invasion) (Ramanathan *et al.*, 2015); vacuolated muscle fibers, and intracellular amyloid protein deposits or tubulofilaments in the cytoplasm or the nucleus (Simons *et al.*, 2016). Pathological manifestations have been historically split between inflammatory and degenerative predictions. Degenerative signs are based on the presence of an abnormal accumulation of β-amyloidogenic (Distad, Amato & Weiss, 2011). These sediments are found in the muscle fibers of sIBM patients, whereas they used to be observed extracellularly in Alzheimer's diseases (Distad, Amato & Weiss, 2011). Moreover, proteins known to be implicated in degenerative processes have been found, such as ubiquitin (Dalakas, 2015). The inflammatory infiltrates in this myopathy include T cells and macrophages as well as, small portions of B cells (Simons *et al.*, 2016).

2.6 Diagnosis and Laboratory Testing

Several imaging techniques can be used in diagnosing and monitoring onset and progression of NAM. Magnetic resonance imaging (MRI) cannot only be essential in locating the distinctive sites of the biopsy but also has ascertained relevance in studying the response rates as the disease treatment proceeds (Dalakas, 2011). PET scanning can also be used in screening patients who have either DM or NAM for incidences of malignancies. MRI is gaining more relevance in the elucidation of IIMs and similarly having great importance in guiding treatment options. Patients can be assessed for availability of edema, inflammation rates atrophy, and adipose infiltration. The procedure

can also help in guiding muscle biopsy location for increased yields (Mandel, Malemud & Askari, 2017). Given that the location of appropriate biopsy sites is directed by the levels of active inflammation compared to others with only chronic alterations or fat infiltration, the biopsies done on the affected muscles are the key to elucidating the nature of the disease (Mandel, Malemud & Askari, 2017). This is because many etiologies are associated with serum CK and may lead to inflammatory changes on MRI of muscle tissue Mandel, Malemud & Askari (2017).

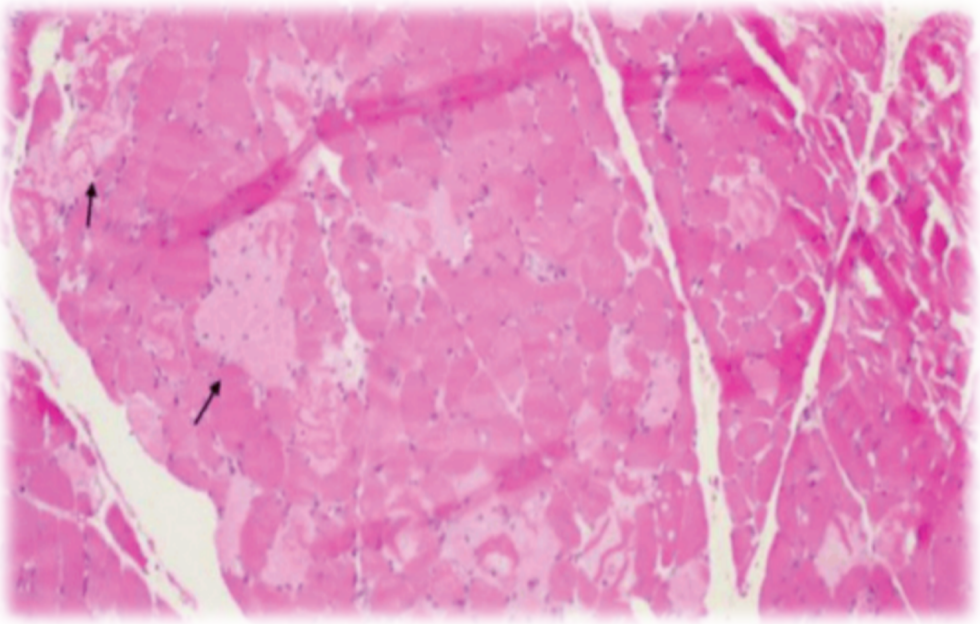

Figure 3: quadriceps muscle biopsy from a patient with necrotizing autoimmune myopathy shows features consistent with necrotizing myopathy without significant inflammation. There are numerous degenerating and regenerating muscle fibers. Some fiber shows necrosis (arrows). (Khattri, S. 2012).

Laboratory testing should be critical in ensuring that responses to treatments as well as disease flares are monitored appropriately as pointed by Mandel, Malemud & Askari (2017). The levels of serum CK can be important in the elucidation of the disease progress by checking the varying elevation rates during treatment even though the initial levels of serum CK can be different among the different subcategories of IIM. Similarly, the elevation of the levels of myoglobin and aldolase can help in the monitoring of the disease progression (Mandel, Malemud & Askari, 2017). Nonetheless, serum glutamic oxaloacetic

transaminase (SGOT) and serum glutamic pyruvic transaminase (SGPT) may be elevated secondary to both liver disease and muscle breakdown (Mandel, Malemud & Askari, 2017). This may render monitoring medication side effects cumbersome especially when treatments used involve methotrexate (MTX) as per the deductions of Mandel, Malemud & Askari (2017). In such an incidence of treatment, gamma-glutamyl transferase level may be needed to separate SGOT/SGPT elevation due to muscle breakdown versus hepatotoxicity (Dalakas, 2015). In this case, thyroid function tests should constantly be done in order to help the patient with thyroid disorders know the progress of their condition (Mandel, Malemud & Askari, 2017).

Electromyography (EMG), as well as nerve conduction studies, have been applied in the separation of separation of neuropathic and myopathic causes weaknesses of the muscles before a biopsy is done and therefore may render the treatment as well as disease progression cumbersome (Mandel, Malemud & Askari, 2017). This method is in most instances performed unilaterally on the upper and lower extremity in IIMs (Dalakas, 2015). It displays a characteristic short duration, low amplitude polyphasic units on voluntary activation with increased spontaneous fibrillations and positive sharp waves (Mandel, Malemud & Askari, 2017).

The detection of myositis autoantibodies in the laboratory is also important in the understanding of the nature of NAM. Single multi-analytic blot test is a good test for determining assaying myositis autoantibodies (MSA/MAA) through screening (Ghirardello *et al.*, 2013). Under this technique, highly purified antigens are attached on the surfaces of nitrocellulose material whereby different samples are absorbed for later analysis. The accuracy of this method highly depends on the biochemical nature as well as the grade of the purified antigens used in this blot-based assay (Ghirardello *et al.*, 2013). The relationship that exists between autoantibodies and myositis indicate that these biological components can be studies and evaluated to determine the progression of a disease as well as the extent of damage caused on the tissues studied (Ghirardello *et al.*, 2013).

More evidence also point out to the fact that autoantibodies linked to myositis may directly correlate to the extent of harm a disease has caused and how fast one can respond to therapy (Ghirardello *et al.*, 2013). Factors worth looking into include anti-ARS, anti-

SRP, anti-MDA5, and anti-HMGCR (Ghirardello *et al.*, 2013). The components appear to be directly linked to both target organ involvement and systemic disease activity, together with biomarkers of myositis in necrotizing myopathies (Ghirardello *et al.*, 2013). The peculiarity in understanding of the cell disease manifestations in NAM has made the diagnosis of NAM difficult. However, the use of autoantibodies that are directly linked to anti-SRP as well as anti-HMGCR is critical in offering guidance as to how the condition can be assayed for and keenly monitored. Currently, the seronegative NAM represents 20-30 % of cases.

According to Orpha.net (2017), diagnosis of NAM is based on visible clinical symptoms and on the screening results displayed after MRI expressing the nature of the muscle. Evidence of minimal or lack of inflammatory infiltrates and marked muscle necrosis are usually a pointer to the existence of NAM,-factors that distinguish it from other inflammatory myopathies as per the deductions on Orpha.net (2017). The procedure based on EMG shows myopathic findings. CK levels are usually more than ten times above the upper limit of normal level at the time of onset of muscle weakness. MRI results may show a variant of patchy edema within muscles (Orpha.net, 2017).

2.7 Treatment and Management

Treatment of the underlying cause of NAM is important (Orpha.net, 2017), and it usually takes care of statin effects as well as malignancies involved. Patients suffering from NAM usually respond to a wide array of treatment options that involve multiple-agent, long-term immunosuppressive therapies beginning with high-dose corticosteroids (Orpha.net, 2017). Although intravenous immunoglobulin (IVIg) appears to be the most effective treatment method, several other methods exist that help in reducing the impact of the condition on one's life. Rituximab has also been shown to have massive positive impacts on NAM treatment. It is, however, critical to note that a treatment plan employed heavily relies on the basis of muscle strength and biologically on the CK levels as posited on Orpha.net (2017).

NAM patients' long-term management of hyperlipidemia has remained a major challenge as per the deductions of Babu & Li (2015). Several methods have however been suggested in managing such a problem. For one, NAM patients should consider avoiding the use of statins completely and consider using other non-statin lipid-lowering agents like ezetimibe or cholestyramine (ACNR, 2017). Other alternatives are related to lifestyle activities such as weight reduction, cessation of smoking, as well as dietary modification (Babu & Li, 2015).

2.7.1 Pharmacotherapy

According to Dalakas (2011), prednisone remains the first-line drug for treating NAM as per the experience rather than drug trials. The general procedure for using this drug is to start with a high dose and slowly adjust to lower doses until the needed efficacy is achieved for the condition. The high dose, in this case, may entail beginning with at least 1mg/kg of 60 to 80 mg per day (Ramanathan *et al.*, 2015). This is a single daily dose taken in the morning after breakfast (Dalakas, 2011). This may be done routinely until 3 to 4 weeks (Dalakas, 2015). After this, the daily dose would be changed to a different routine from the first one as guided by the efficacy state and side effects until the lowest possible dose that controls the disease is ascertained (Dalakas, 2011). However, there is a difference in the case whereby respective patients have the severity of infections. In such instances, the treatment may begin with intravenous methylprednisolone (1g/day or 3-5 days) and then be switched to the oral regimen (Dalakas, 2011). Immunosuppressant may also be added at the outset even though this may create some uncertainty regarding the value of the added drug in the long run as explained by Dalakas (2011).

Some of the drugs have been listed to have some effects in treating NAM. The first one is Azathioprine, which is an antipurine antimetabolite and used in doses of up to 3mg/kg (Dalakas, 2011). This drug takes six months to reflect positive outcomes hence it has been falling out of preferences for most medical suggestions. Second, to this, Mycophenolate mofetil (Cellcept) is also an antipurine antimetabolite but is preferable than azathioprine (Danieli *et al.*, 2016). Given that it works faster than the first drug, it is more expensive to acquire. The dose is taken twice a day and involves 2-3 grams

(Dalakas, 2011). Under instances of organ rejection, the drug works faster by inhibiting the production of new B and T cells. It can also be applied in conjunction with other drugs such as prednisone in treating autoimmune disease by initially being applied to autoreactive, existing lymphocytes. It is also effective in treating dermatomyositis in small cases as indicated by Dalakas (2011).

Methotrexate is an antagonist of folate metabolism (Danieli *et al.*, 2016); and is also important in the treatment of NAM. The drug is given orally beginning with a dose of 7.5mg on a weekly basis for the first three weeks (with daily doses given thrice constituting 2.5mg after every 12 hours) (Dalakas, 2011). The dose is usually increased gradually to 2.5 mg per week up to a total of 15-20mg weekly (Dalakas 2015). Since the dose works faster than azathioprine, it is preferred by some medical practitioners as a steroid-sparing agent (Dalakas, 2015). Nonetheless, the drug is less expensive than CellCept but more toxic (Ramanathan *et al.*, 2015). Some of the underlying negative outcomes of the drug include stomatitis, hepatotoxicity, as well as other gastrointestinal symptoms (Dalakas, 2015).

Fourthly, cyclosporine is also another drug of utmost relevance in treating NAM. The drug is given twice a day with doses adding up to 150mg. Cyclosporine acts faster than azathioprine and Cellcept, but the levels of toxicity are higher hence calling for regular monitoring of the renal function and blood counts (Dalakas, 2011). This type of drug is just but one of the various options available for treating NAM. Apart from discontinuing a drug with adverse side effects in the body like statins, immunosuppressive therapy should be immediately adopted as it forms the backbone of treatment mechanisms underlying this condition as asserted by Babu & Li (2015). Some patients with statin-induced NAM only require the cessation of the drug use as they respond positively to such stoppages as outlined by Mammen (2016). Other forms of therapy should only be adopted in cases whereby the cessation of statin use does not trigger substantive outcome for the patients. In case discontinuation of statin therapy does not mean well for the patient, the statin use should be stopped and the patient treated with other immunosuppressive forms as applied to other patients who have other forms of muscle diseases (Mammen, 2016).

Prednisone together with or without methotrexate is usually the first preference on treatment even though the use of azathioprine, cyclosporine, intravenous immunoglobulin,

mycophenolate, as well as rituximab are also used in different combinations (Babu & Li, 2015). According to Advances in Clinical Neuroscience and Rehabilitation (ACNR) (2017), the treatments for patients with NAM is slowly shifting towards early aggressive immunotherapy especially with subgroups that test positive for the presence myositis autoantibodies. This then follows that the treatment of NAM with more than one form of immunotherapy immediately after diagnosis has a chance of yielding a positive outcome in the first three months of disease diagnosis (ACNR, 2017). However, the use of corticosteroids is highly influenced by the time of diagnosis of the condition with doses influenced by clinical and biological responses as asserted by ACNR (2017). Not all patients would effectively respond to steroid-sparing medications such as methotrexate, mycophenolate, and azathioprine; some patients relapse to weaning, calling for the use of an additional agent such as intravenous immunoglobulin g (IVIg), Rituximab, or Cyclosporine (ACNR, 2017).

Even after the NAM patients have fully recovered their strengths, immunosuppressive medications should be maintained in order to ensure that certain symptoms do not return and keeping in mind that some patients may face relapse hence calling for uninterrupted treatment process until there is full certainty that the patients have fully recovered as cited by Mammen (2016). It is also worth noting that some patients may regain their full strengths even though their CK levels may still be elevated, suggesting that the process of muscle regeneration may be faster than muscle destruction. Similarly, in some cases, muscle weakness may still prevail even when the muscle enzyme levels of the patient have already returned to normal as cited by Mammen (2016). This occurs in situations that entail long-term under-treatment of NAM hence a permanent damage leading to the replacement of muscle tissue ensues as per the deductions of Mammen (2016). Patients with a statin-induced form of NAM have high chances of recovering well from the condition if the investigation is made quick enough and appropriate actions taken. Fast discontinuation of statin use is key to ensure that muscle damage does not turn out to be severe. The treatments should follow the rightful procedures, and MRI should be used in guiding the medical practitioners on the extent of damage already caused to the muscle. Good patient outcomes are highly dependent on the time of diagnosis and the drugs applied following the failure of the first line of

immunosuppressive therapy or after cessation of statin use in cases of statin-induced necrotizing autoimmune myopathy.

The second line of pharmacotherapy involves the application of immunoglobulin treatments intravenously. This method is usually applied as the second treatment option when the first-line treatments involving corticosteroids have failed to yield the desired impact. In the treatment of NAM, the desirable impacts of IVIg have epidemiological documentation, but as per the experiences recorded from patients, it can lead to great relief to respective users as posited by Dalakas (2011). The intravenous application is beneficial for diverse mechanisms of actions, including barring complement activation, limiting binding of autoantibodies on muscle cells, alteration of receptor binding on macrophage surfaces, cytokine inactivation, or alteration of antigen recognition by activated T cells (Dalakas 2015). Patients that have shown poor response to the drugs are liable for reassessment and further diagnosis in order to ascertain the real cause of such poor responses completely. Factors that are worth looking at and evaluating include the patient's clinical history, the pattern of an individual's muscle weakness, muscle biopsy findings, as well as family history as cited by Dalakas (2011). It is also necessary to perform a new biopsy in order to bolster confidence in the reports obtained following the re-evaluation of previous poor response.

Confirmation of diagnosis permits the use of the following biological components: Cyclophosphamide; calcineurin phosphatase activity inhibitors; and new cellular components in entailing fusion proteins or monoclonal antibodies (Dalakas 2011). These biological components target B cells and T cells. They are also capable of aiding in T cell migration, prevention of complement formation, TNF-a strategies, as well as cellular adhesion as posited by Dalakas (2011). All these factors brought together are essential in ensuring that the drugs used by a patient are effective. Also, the cellular components are key to ensuring that muscle recovery process if fast.

Corticosteroids

The first-line of therapy in the treatment of NAM is the use of corticosteroids. In this case, either prednisone or prednisolone is used to treat the patient. Even though both oral and intravenous methods of application of these medications exist, oral

dexamethasone pulse is superior to oral prednisone and so is intravenous methylprednisolone to oral prednisolone as cited by Dalakas (2011). If the medications do not trigger any positive changes by the third month, then an alternative way of treatment is adopted that involves the use of steroid-sparing regimes (Dalakas 2011). This form of treatment is highly effective when the where muscle damage is rarely adverse.

Although there are no controlled trials of corticosteroids, there is general agreement that they are effective in DM, PM, and NM. Corticosteroids can be used in a wide range of regimens and routes of administration. Prednisone 1 mg/kg/d (60–100 mg) is often given for 4 weeks followed by an abrupt or tapered conversion to an every other day schedule. A course of intravenous methylprednisolone may be administered first in patients with severe weakness. This taper is slower in patients with severe disease. A daily corticosteroid schedule is necessary in well-controlled hypertensive or nonbrittle diabetic patients. Although many patients feel immediately better after starting corticosteroids, strength improves over 2 to 3 months.

Methotrexate

Methotrexate (MTX), an antifolate that inhibits lymphocyte proliferation, is an effective and rapidly acting second-line steroid-sparing immunosuppressant. Methotrexate is started at a dose of 7.5 mg taken once weekly. The dose is increased 2weeks later to 15mg/week in two divided doses. The dose is then increased by 2.5 mg per week at 3 months, depending on the clinical response. The maximum weekly dose is 25 mg. Folic acid 0.8 to 1 mg per day orally is given to prevent stomatitis. Some advocate holding folic acid on the day of MTX administration. (Castro, 2012).

2.7.2 Physiotherapy

Several supportive and assistive measures are useful in limiting limb contractures, respiratory compromise, swallowing and gastrointestinal issues, and other complications that develop secondary to the weakness associated with most myopathies. The judicious implementation and monitoring of these measures is a crucial component to the management of myopathic patients.

According to Barsotti *et al.* (2015), physical training is one of the best ways to improve cardiorespiratory function as well as strength. Regular exercise and training can aid in the elimination of some NAM symptoms such as fatigue, pain, and muscle weakness that lead to low life quality of an individual that suffers from the condition. As outlined by Barsotti *et al.* (2015), training to bolster one's strength or aerobic-based exercise programs in individuals having a myopathy might enhance muscle strength; imparts heart/respiratory functions and prevents disuse atrophy (Barsotti *et al.*, 2015). A study has also indicated that training, even though had no significant effect on the level of CK in the affected individuals, led to an improved heart rate that was lowered, no muscle damage during the exercise period hence citing positive progress in heart-related fitness (Barsotti *et al.*, 2015). The deductions of such a project indicate that aerobic training is key to strengthening the heart rates in persons who have necrotizing autoimmune myopathy (NAM) (Barsotti *et al.*, 2015). Even though exercise is important for the improvement of muscle performance and health in NAM patients, it is important to evaluate the impact that such physical activities might have on active disease (Barsotti *et al.*, 2015). This implies that physical exercise should be considered with utmost caution as a way of improving on the recovery of the damaged muscles.

Patients with muscle disorders invariably ask whether exercise will improve their strength, and it is not uncommon for patients to begin intense exercise programs soon after diagnosis. While the concept that exercise can improve even damaged muscle is appealing, there is a theoretical risk that exercise may increase muscle damage, especially in the inflammatory myopathies and genetic disorders affecting structural proteins (eg, dystrophin in DMD). However, exercising has significant benefits beyond effects on strength, such as maximizing cardiopulmonary function, limiting osteoporosis, and improving patients' well-being.

A wide array of systemic clinical indications that most regularly happen in patients with NAM include but not restricted to shortness of breath, fatigue, and muscle weakness (Physio-pedia.com 2017). In such cases, the most affected body parts are the musculature proximal to the trunk with weaknesses reported mostly in the neck, back, shoulders, forearms, thighs, as well as hips as indicated on Physio-pedia.com (2017). Even though distal weaknesses may also occur in the region, they are less common, and the trunk is usually left strong as compared to when the proximal muscles are the ones affected. Besides the above-affected parts, respiratory musculature is also affected hence calling for the adoption of both resistance and aerobic training as ways to counter such effects (Physio-pedia.com, 2017).

Several schools of thought posit that exercise is beneficial to patients under various categories of sickness, helping those with chronic conditions, stable conditions, as well as those that have had active and reset onset of NAM (Physio-pedia.com, 2017). It is however advisory for patients under acute exacerbations to utilize pharmacological management in control their inflammation rates before starting active exercise program; they should also limit their physical activities to normal functional mobility in order to bolster faster muscle recovery that does not extend the already existing nature of harm on the patients (Jain & Sharma, 2014). The main focus on physical therapy as a way of instilling recovery and management of NAM patients is to ensure that normal body functions are maintained, and fall risks are tremendously reduced. In order for the patients to maintain the function of their body parts, they should maintain exercise, and this should be done for about 5-6 days on a weekly basis (Physio-pedia.com, 2017). However, exercises that entail the strengthening of the body should not occur consecutively within a week as patients are advised to consider active resting days when they focus only on positioning and relaxation rather than on strengthening as cited on Physo-pedia.com (2017).

Chronic or progressive muscle weakness often leads to soft-tissue contractures. These are caused by a complex set of factors including muscle strength imbalance, prolonged immobility, the effects of gravity, and other factors inherent in the disease pathogenesis. For example, hip, knee, and ankle joint contractures are common in nonambulatory patients and are especially difficult to prevent if the hip flexors, knee

flexors, and ankle plantar flexors remain stronger than the corresponding muscle antagonists. There are some theraputic aproaches to avoid that, mainly stretching and positioning. Although most clinicians and therapists recommend regular stretching for patients with chronic myopathies, no firm evidence has shown that soft-tissue joint contractures can be prevented even by aggressive positioning, range-of-motion exercises, or the use of night splints.(M. King, 2013).

Physical and occupational therapy, orthotic devices, and exercise are important components of myopathies therapy, as early as 2 to 3 weeks from the acute phase. Although other studies have reported the safety and benefits of resistive exercise in active patients 1 to 3 months into their treatment, most of the studies have been in chronic PM or NAM. In severe cases, passive range of motion exercises are prescribed for 1 to 3 months or until strength and CK start to improve, at which point strengthening exercises are initiated. In patients with mild to moderate weakness, a strengthening program is started 2 to 4 weeks after steroid initiation. Because pain from arthralgia and possibly arthritis is relieved by joint flexion, early mobilization is important to prevent flexion contractures of large and small joints, especially in juvenile dermatomyositis (JDM).(Castro, 2012).

Aquatic therapy is an essential component of physical therapy for patients with NAM. This type of therapy is of utmost relevance in improving the quality of life of an individual since it aides in several aspects. For one, it helps in acquiring therapeutic benefits without exposing one to fall risks. Secondly, the turbulence in water is critical in assist in blood flow to the affected skin surfaces. Thirdly, buoyancy helps in the acquisition of stable posture and reduces extreme muscle weakness. Water resistance can also be control through modified movements of the patient. Last but not least, this type of exercise provides the ability to mirror functional movements with decreased energy expenditure (Physio-pedia.com, 2017). Exercising in this manner, therefore, offers a myriad of benefits to NAM patients that other physical activities alone like walking cannot offer hence highly recommended in bolstering fast recovery and improved quality of life.

In the past, patients with neuromuscular diseases were advised not to exercise because of the fear that too much exercise might produce "overuse weakness." No controlled studies have demonstrated that the phenomenon of overuse weakness actually

exists. Most studies of exercise training in patients with neuromuscular disease, despite methodologic limitations, suggest that strength and aerobic capacity gains can occur in patients with slowly progressive disorders. Four forms of exercise training are relevant to patients with neuromuscular disorders: flexibility, strengthening, aerobic, and balance exercises. (Krivickas, 2003).

2.7.3 Occupational Therapy

Occupational therapy is an important aspect of recovery and disease management for individuals that suffer from NAM. It is therefore critical for organizations to create an effective working environment for employees that have the condition to adjust effectively to the workplace environment. It is also prudent for employees that are recovering from NAM to be treated with the utmost dignity they deserve in order to instill into them a sense of belonging. This is important in promoting the stability of their emotional as well as psychological well-being hence leading to fast recovery. The role of occupational therapy should be regarded with utmost relevance since it offers patients an opportunity to continue with their normal lives and at the same time gradually recover from the ailment in a good working environment with ease. Workers that have NAM and are recovering hence the need to return to work should be offered with the necessary support within respective workplaces so that their recoveries do not take beyond normal duration on recovery as well as helping them prevent incidences of symptom relapses.

In order for an organization to reduce poor production outcomes due to factors such as employee back pains, it has to set up facilities and services that help in workforce adjustment such as those with NAM as pointed out by Leyshon & Shaw (2008). Presenteeism is a term that refers to low productivity that is realized within an organization due to poor productivity from the present employees as a result of factors such as fatigue, illness, as well as injury (Leyshon & Shaw, 2008). With many organizations indicating that this factor has more detrimental impacts than absenteeism, it

is important for respective organizations to ensure that such statistics do not affect their work as opportunities for good work delivery are put in place for those employees that may be adjusted to returning to work following an ailment such as NAM. The price that is paid by the corporate organizations, as well as the societies from which NAM patients come from, are higher as compared to when conducive environments could be created both within societies and in workplace environments for helping such individuals with their recovery plans as intended by physicians. The prices are paid in terms of financial costs, pain, suffering, as well as low life qualities as cited by Leyshon & Shaw (2008).

Several ways exist through which workers that are recovering from NAM can be helped to adjust at the workplace. Application of ergonomics at the workplace is important as cited by Leyshon & Shaw (2008). This method entails the study and the process of designing and modifying tools, equipment, materials, workplaces, tasks, jobs, merchandises, systems, as well as environments to suit the abilities, limitations, plus societal needs of humans in the workplace (Leyshon & Shaw, 2008). This principle is important in that it fuses all the aspects that can lead to effective adaptation as well as recovery at workplace into one major goal. Ergonomics is important in instilling occupational therapy since all the necessary tools needed for one's recovery are provided with ease in order to bolster diversity that exists within an organization. The issues addressed by this technique encompass the entire workplace environmental issues such as policies, attitudes, and policies in a bid to bolster inclusivity (Leyshon & Shaw, 2008).

In most cases, the ideas presented under ergonomics are guided by the core principles of ethics that include the principle of common good. This principle demands that all the activities within any organization be for the good of all the people including the workers and the external community members. Treating all workers fairly and ethically helps to inculcate a sense of belonging and an assurance that one is respected and is counted on as an important driver for change within such corporate entities. Issues of satisfaction at the workplace are key to determining how one adjusts to recovery guidelines provided by respective health practitioners. It, therefore, follows that how an organization handles its workers that are returning to work following an ailment has much weight on the performance of the very organization as well as impacting the recovery of an individual. Workers should be treated ethically based on all aspects of life such as

behavioral aspects, physical factors and emotional attributes alike as asserted by Leyshon & Shaw (2008). The regulations are also related to the governmental policies such as protection of worker rights via the labor department, and provision of insurance policies that enable workers to seek any immediate medical services in case unexpected symptoms are experienced or complete relapse occurs.

Workplace setting plays a key role in aiding patients with recovery. The equipment or machines that are set in place should appropriately align to a returning worker's cognitive, physical as well as social traits (Leyshon & Shaw, 2008). Some of the adjustments that can help workers with occupational therapy include adapted computer keyboards and office chairs; reorganized work station that takes care of the nature of the worker; creating opportunities for job task rotations; or even personalizing a work station set-up or piece of equipment by the worker (Leyshon & Shaw, 2008). Such activities have the highest chances of reducing injury rates in workers that still have the massive task of adjusting to the gradually ending symptoms of NAM.

It is worth noting that education also plays a major role in helping NAM patients recover well within their work places. The education given to returning workers should involve insights such as keen guidelines on performing specific tasks to improve body stability, gait as well as how to learn using body parts to perform work demands in divergent ways. This would be important in helping to stretch some moving body parts as a way of exercise while at the same time working as pointed out by Leyshon & Shaw (2008). Organizations should also consider providing the returning workers that suffer from NAM with sit-stand stools so that sitting and standing should be alternated as needed without causing much harm to the recovering worker. There is also need to give such workers mini-breaks that allows them to stretch and apply the necessary treatment guidelines as needed. Effective occupational therapy at workplace therefore highly relies on how an organization's management conforms to the ergonomics that ensure that standards are inculcated on employees' return to work procedures. Communication at workplace is also key as organizations should embrace communication structures and strategies that ensure that all workers understand what is expected of them at workplace with utmost ease required.

2.7.4 Prognosis

As pointed out by Danieli *et al.* (2016), NAM being one of the IIMs is associated with increased mortality rate. The disease is also associated with affecting other organs and causing diseases such as cardiovascular and lung disease, with the latter being pronounced as the being factor leading to high mortality rates. The rates of the mortalities due to cardiovascular attacks could be very high even the existing epidemiological data are not being representative of the actual fatalities. Cardiac related diseases have been on the rise in patients suffering from NAM, and these have led to the loss of lives due to cardiac arrests, heart failure, myocardial infarction, as well as, arrhythmia as cited by Danieli *et al.* (2016). Some studies have also pointed out to the fact that the association of gender, heart involvement, interstitial lung disease, and arthritis have increased the mortality rates (Danieli *et al.*, 2016). Moreover, over the years the male gender has suffered more mortalities (Danieli eta l., 2016). Given that approximately 70% of NAM patients only have subclinical manifestations of the symptoms highlighted herein, the prognosis of the disease is regarded as being severe as it involves features that are divergent and life threatening if not handled in good time by both the patient and the medical practitioners (Danieli *et al.*, 2016). It is therefore essential for medical practitioners to evaluate the cardiac performance of a patient that has been diagnosed by NAM routinely in order to reduce the unintended harms that may arise as a result of such manifestations. Given the underlying risks, it is important for patients to be routinely screened through MRI to assess the impact of damage to the muscles as well as elucidating the existing risks as a result of the ailment's relationship to diseases of the heart.

According to Nichols *et al.* (2015), early treatments are important in reducing muscle atrophy as well as loss of functionality. Besides mortalities being linked to cardiovascular and lung conditions, some death incidences are also as a result of cancer cases. Therapy initiation works for the better improvement of patients' lives especially when the patients involved have a younger age and also if the manifestations have been elucidated and therapy adopted with immediate effect as Nichols *et al.* (2015) outlines. Negative aspects of prognosis are in most cases related to older age of onset, pulmonary complications, esophageal involvement, as well as the above mentioned- cancer. This

implies that the severity of NAM highly depends on the age of onset as well as the age of the person at the point when one is diagnosed. Patients that are diagnosed early enough in their tender ages have high chances of recovering well as continuing with their normal lifestyles after having gained the needed quality of life.

On the other hand, patients that have been diagnosed late in their old ages have chances of surviving as well. However, this can effectively work in case the disease is diagnosed at its early stages and appropriate measures taken immediately to counter the muscle damages. Age also is consider as an important factor since the immunity of one decreases with age. This implies that the amount of time needed for a person at a tender age to recover is different from the duration it would take an old person to fully recover and regain the desirable quality of life. It is therefore important for patients with NAM to reduce the underlying risks by constantly going for screening procedures that are focused on determining the presence of other associated conditions like cardiovascular, cancer, lung disease, as well as pulmonary complications if any. This would positively enhance the treatment processes since any chances of underlying risks would be handled before they have permanent effects on an individual's life ultimately leading to death.

To sum up, necrotizing autoimmune myopathy is a condition that has not been fully evaluated in terms of the epidemiology and existing medical management plans. However, more research should be done on how to help patients that have the condition to fully recover with ease and reduced pain. In order to ensure that the mortalities resulting from the disease are reduced, patients should adopt the art of constant screening such that all the underlying risks can be noted in good time. Therapy should be adhered to without hesitation as it is one way through which the condition can be managed. Nonetheless, both physical therapy as well as occupational therapy play a crucial role in the recovery of patients suffering from NAM. Strict adherences to therapies are essential in reducing symptom relapses, pain, as well as the recovery time hence leading to the overall improvement in the quality of life. The role of both the society as well as the workplaces that individuals having NAM come from is also important. Fellow employees should always support such colleagues in their times of need by sharing with them how certain tasks are done to help them adopt to new lifestyle changes following the injuries caused by the disease. The management should also ensure that occupational therapy facilities

and equipment are in place so that they abide by the ethical codes of corporate leadership and governance.

The support of the government is needed in ensuring that services such as MRI and EMG are readily available for patients. Making such services readily available is important since it would cut on the costs for individuals to check on the nature of disease progression. Supporting such initiatives as well as raising public awareness for people to go for screening is essential in reducing the mortality rates of NAM. Given that a lot of epidemiological data is lacking in terms of the severity of the condition and the effective therapeutic ways to manage it, more research should be done on the already existing large gaps so that information can be available necessary to guide both medical practitioners and patients to this condition- NAM.

3 Special Part (Case Study)

3.1 Work Methodology

I have done my bachelor's practice in rehabilitation centre in Rehumatology institute in Prague, from 30. 1. 2017 to 10. 2. 2017 my work was guided and supervised by Bc. Petr Velišek.

My thesis patient is a woman who had necrotizing autoimmune myopathy. The patient has been informed about my thesis practice so we could cooperate and that her personal information, anamnesis and her present situation will be used

7 therapeutic sessions I spent with the patient every morning. Initial and final examinations were included in beginning of first session and at the end of last sessions respectively.

I used a measuring tape, plastic goniometry during both initial and final examinations. Softball, medicine ball and spongy balls were used during the therapy sessions.

However, according to the instructions from the supervisor, therapy procedure should mainly focus on neck and upper extremities, so we were concentrating on it by using strengthening and improving range of motion, PIR according to Karel Lewit and PSF according to Janda were also included in the therapy sessions.

3.2 Anamnesis

Examined person: A. M

Gender: Female

Year of birth: 1955

Diagnosis: Necrotizing autoimmune Myopathy (NAM)

STATUS PRAESENS:

Subjective:

The patient doesn't feel pain but rapid exhaustion and hardness of breathing after short time effort or any physical activity. Ability to walk with crutches 2 minutes. The patient feels weakness of whole body muscles as well as dizziness during verticalization.

Objective:

There is significant weakness of all body muscles especially right upper extremity and left lower extremity. Weakness of most of body muscles due to lack of movements and bed hospitalization for many years. Hypotonic muscles are clear of lower extremity especially area of ankle joint in both legs. The patient is unable to keep the head in neutral position during standing or walking. There is no limitation of hand functions so the patient is able to eat and drink without assistance as well as using the crutches. She is able to walk using crutches around the room for maximum 3 minutes then she will be completely exhausted. The patient has no problem of breathing during static phase but due to effort or exercising she will be tired then breathing hardening gradually.

Assistive devices: Crutches while walking

Dominant limb: Right limb

Glasses: only while reading

Height: 158cm

Weight: 82 kg

BMI : 27.1

Chief complaint:

Significant weakness of whole body muscles especially neck flexors, dropped head and rapid exhaustion. The patient doesn't complain of pain.

History of the case:

The patient is 62 years woman has been suffering from autoimmune disease, which is necrotizing autoimmune Myopathy for over 8 years. She has been hospitalized for over than 7 years in different clinics and hospitals. She was diagnosed with myopathy in December of 2009 for that was the first hospitalization in Pardubice for one week; she was under medication only, without any physical training. The patient's body started to reject any medication in 2013 therefor the case was worsening gradually in the beginning of 2014 the patient was on bed, excessive fatigue and muscles weakness combine with The loss of ability to walk. After that the patient was sent to Revmatology Ustáv for better rehabilitation. She spent almost 2 months for examinations and check up in RÚ. The doctor had to find out a substitution for her medication, after finding a drug which her get along with. Since then until the end of 2015 the patient was getting better by both the medication and physical therapy, in 2016 the patient is exercising regularly, she is able to sit and stand up without assistance and she is able to walk with crutches around the corridor usually not more than 3 minutes.

Diet: no diet

Functional anamnesis/history:

The patient struggles the most when she wears or takes off her clothes, the decreased of her left hand's function limits some activities such as eating, driving, fixing things etc.

Family anamnesis:

The patient's mother suffered from Gastritis, and her brother was diagnosed with multiple sclerosis (MS). The patient has one healthy child born in 1988 (29).

Occupation anamnesis:

The patient worked as store manager until November 2009.

Social anamnesis:

She lives with her husband in the second floor -17 stairs –

Sport, regular physical activity:

None.

Pharmacological anamnesis:

Pulsy Solu- Medrolu 5g, Prednison 70mg at the beginning then decreased the dose to 50mg, Methotrexate 2,5 mg, Azathioprin 150mg, Cyklosporin 150mg, Lusopres 20mg, Equuoral 50mg.

Abuses: Alcohol occasionally and coffee every morning, she doesn't smoke.

Past operations: Appendectomy (2009) and hysterectomy (2010).

Prior Rehabilitation:

I. 2010-2013 Neurology department in Pardubice:

The patient was diagnosed with myopathy unknown etiology with drop head, patient was in bed. Medications were prescribed for the patient without physical exercises due to patient status at that moment: (Pulsy Solu-Medrolu 5g in total), (Prednison 70mg). The patient was under medication for almost year and half then she started to feel better, she could transfer from bed to sitting on chair with assistance. So physical exercises were applied such as active and passive movements, breathing exercise and strengthening exercises.

II. 5.2014: Hospitalization in Revmatologicky Ustáv:

The patient was completely lying on bed with some side effect of medication she used to take before, her body started to reject the drug and her statues was worsening rapidly. So the doctor was looking for better substitution for medication. Methotrexate was prescribed for the patient and her body started to reach properly to this medication.

No physical exercises were done due to patient status.

III. 2.2015 - 5.2015: Hospitalization in Revmatologicky Ustáv:

The patient didn't feel pain that time, so physical exercises were recommended to the patient.

Physical examination was done by the therapist and found out: blockage of left wrist and metacarpal joints, weakness of body muscles in all segments, hypotonic muscles in extremities, right lower limb and left upper limb in better condition.

Physical exercises were focused mainly in strengthening the extremities and upright the body position. After almost 3 months of rehabilitation, the patient couldn't tolerate the whole therapy session that's why the therapy decreased to be done 15 minutes twice a day, morning and afternoon.

There was no such improvement in her case except the hand functions got improved in and transferring from bed to stand up becomes maturated and faster.

IV. 2016: Hospitalization in Revmatologicky Ustáv:

The patient didn't complain any pain; weakness of whole body muscles, hypotonic muscles in extremities and fatigue in all body segments after any exercise or small effort. The patient was exercising daily; the patient went under many exercises such as passive and active exercises, strengthening exercises, also verticalization education, the patient felt dizziness after verticalization.

The patient is able to sit on bed and stand up but unstable, walking with crutches. In general the patient case is getting better.

3.3 Initial Kinesiological Examinations By Physiotherapist:

The examination is done on Tuesday 33th, January 2017

3.3.1 Postural Examination-Aspection-Plumb Line Test:

The patient is able to stand up next to bed for 1-2 minutes then she will be tired. She has difficulty of upright the head position.

Back View	
The base of support	Narrow
Shape of the heels	Valgus heel
Position of the ankle joints	Valgus
The calf muscles	Hypo-tone symmetrically
Position of the knee joints	Valgus Knees
Position of pelvis	Elevation of right side
Paravertebral muscles	Hypo-tone
Curvature of the spine	Physiological curvature.
Position of the scapula	Elevation of left scapula
Position of the shoulder girdle	Elevation of left shoulder
Position of head	Dropped head forward

Table 1 : Initial Aspection of back view

Side View	
Position of the ankle joint	
Position of the knee joints	Left knee is forward
Thigh muscles	Hypo-tone both sides

Curvature of the spine	Physiological curvature in Thoracic spine and flat lumbar, hyper lordosis in cervical spine.
Position of shoulder joint	Left shoulder is higher
Position of the head	Head is dropped forward

Table 2: Initial Aspection of side view

Front View	
Weight distribution	Left tendency
Position of the knee joints	Valgus
Shape of the thigh muscles	Hypo-tone both sides
Position of the pelvis	Elevation of right side
Position of the shoulder girdle	Elevation of left shoulder
Position of the head	Protraction

Table 3: Initial Aspection of Front view

3.3.2 Goniometry -SFTR-:

Hip Joint				
	Active		Passive	
	Left	Right	Left	Right
S	10° - 0 - 70°	10° - 0 - 70°	10° - 0 - 90°	10° - 0 - 110°
F	30° - 0 - 10°	40° - 0 - 10°	60° - 0 - 30°	60° - 0 - 30°
R	40° - 0 - 30°	40° - 0 - 30°	50° - 0 - 40°	50° - 0 - 45°

Knee Joint				
S	0° - 0 - 90°	0° - 0 - 90°	0° - 0 - 110°	0° - 0 - 120°
Ankle Joint				
S	20° - 0 - 40°	25° - 0 - 40°	30° - 0 - 50°	35° - 0 - 50°
R	10° - 0 - 30°	15° - 0 - 35°	20° - 0 - 40°	20° - 0 - 40°

Table 4: Initial Goniometry of lower extremities (degree).

Shoulder Joint				
	Active		Passive	
	Left	Right	Left	Right
S	10° - 0 - 80°	30° - 0 - 90°	30° - 0 - 140°	50° - 0 - 160°
F	80° - 0 - 0°	90° - 0 - 0°	130° - 0 - 0°	160° - 0 - 0°
T	90° - 0 - 15°	90° - 0 - 20°	110° - 0 - 30°	120° - 0 - 30°
R	75° - 0 - 65°	75° - 0 - 65°	70°- 0 - 65°	80° - 0 - 70°
Elbow joint				
S	0° - 0 - 110°	0° - 0 - 100°	0° - 0 - 120°	0° - 0 - 120°
Wrist Joint				
S	70° - 0 - 70°	70° - 0 - 80°	75° - 0 - 80°	80° - 0 - 80°
F	15° - 0 - 20°	20° - 0 - 30°	20° - 0 - 30°	20° - 0 - 30°

Table 5: Initial Goniometry of upper extremities (degree)

3.3.3 Gait Analysis:

The patient is able to walk around the room with crutches. Drop head during walking with slight rotation of pelvis. The patient has duck walk due to weak hip muscles.

Base of support	Narrow

Walking rhythm	Non Periodic
Walking speed	Slow
Stride length (short steps/long steps)	Short step
Movement of the foot	Absence of heel strike
Axial position of the lower limb	Flat Foot (both)
Position and movements of the pelvis	Slight swinging
Movement of center of gravity (COG)	Not stable
Position and movements of the trunk	No rotation due to crutches
Activity of back and abdomen muscles	Slightly
Position and movements of the head	Protraction. * It is difficult of up righting the head position during walking
Movements of the upper extremity	Synchronicity *holding crutches;

Table 6: Initial Gait analysis

3.3.4 Specific Testing

- **Romberg Test** (I-III): the patient was able to do the test I- without losing her balance but she lost her balance while doing Romberg II-III.
  Romberg I – Negative
  Romberg II – Positive
  Romberg III – Positive

- **Modification of Standing:**
  Standing on tiptoes: Positive.
  Standing on heels: Positive.

3.3.5 Anthropometric Measurement (length, circumferences)

Upper Extremities	**Left**	**Right**
Length of upper limbs	65 cm	66 cm

Length of humerus	25 cm	25 cm
Length of forearm	24 cm	24 cm
Length of hands	17 cm	17 cm
Circumference of upper limb	39 cm	39 cm
Circumference of forearm	35 cm	35 cm

Table 7: Initial Anthropometric measurement (upper extremities)(cm)

Lower Extremities	**Left**	**Right**
Anatomical length	97 cm	97 cm
Functional length	83 cm	83 cm
Length of thigh	40 cm	40 cm
Length of middle leg	32 cm	32 cm
Circumference of thigh	78 cm	78 cm
Circumference of knee	65 cm	65 cm
Circumference of calf	62 cm	62 cm
Circumference of ankle	47 cm	47 cm
Circumference of foot	21 cm	21 cm

Table 8: Initial Anthropometric measurement (lower extremities) (cm)

3.3.6 Spine Distances:

Test	Result
Thomayer	+16 cm
Stibor	7 cm
Shobar	4 cm
Forest Fleche	5 cm
Lateral flexion	20 cm symmetrical

Table 9: Spine distances (cm)

3.3.7 Muscles Length Test:

According to Kendall 2005
According to Janda 2013

Muscles	JANDA		KENDAL		Result
	L	R	L	R	
Plantar flexors	1	1			Shortness with 5° of dorsal flexion
Hip flexors			>80°		Marked shortness of one and two joints of hip flexors.
Hip adductors	1	1			Shortness of both hip adductors with more limitation of left side
Hamstring			70°		Slight shortness of both hamstrings with 70°
Paravertebral Muscles	2				Marked shortness of paravertebral muscles
Pectoralis major- Upper Lower			90° 135°		Normal length of both upper Pectoralis Shortness of both lower Pectoralis
Pectoralis minor					Shortness was found in both sides.
Levator scapula	1	1			Moderate shortness Levator scapulae
Trapezius (cranial)	2	2			Hard barrier was found
Sternocleidomastoid	1	-			Soft barrier was found while examine the left side.

Table 10: Initial Muscles Length Test.

3.3.8 Muscles Strength Test: (According to Kendall)

Tested Muscle	Left	Right
Soleus	3	3
Gastrocnemius	3	3

Palntaris	3	3
Hamstring	-3	3
Biceps femoris	-3	3
Hip flexors	3	+3
Tensor fascia latae	-3	-3
Gluteus minimus	-3	-3
Gluteus medius	-3	-3
Gluteus maximus	3	3
Hip Adductors	3	3
Neck flexors	2	2
Neck flexion with rotation	2	2
Neck extensor	2	2
Deltoid Medially	3	3
Anterior deltoid	3	3
Posterior deltoid	3	3
Teres major	3	3
Pectoralis major, upper part	3	3
Pectoralis major, lower part	3	3
Pectoralis minor	-3	-3
Biceps brachii	-4	-4
Triceps brachii	4	4
Latissimus dorsi	3	3

Infraspinatus	3	3
Levator scapulae	3	3
Trapezius (cranially)	3	3
Rectus abdominals	2	

Table 11: Initial Muscles strength test.

3.3.9 Muscles Tonicity:

Tested Muscles	Palpation	Position
Levator scapula	Hypotonic in right side with no pain	Supine
Trapezius	Hypotonic in right side with no pain	Prone
Sternocleidomastoid	Hypotonic in both sides with no pain	Prone
Supraspinatus	Hypotonic in both sides with no pain	Prone
Infraspinatus	Hypotonic in both sides with no pain	Prone
Pectoralis major	Normal tone in both sides without pain	Prone
Pectoralis minor	Normal tone in both sides without pain	Prone
Latissimus dorsi	Hypotonic in both sides with no pain	Prone
Deltoid	Hypotonic in both sides with no pain	Prone
Biceps brachii	Normal tone in both sides without pain	Supine
Triceps brachii	Normal tone in both sides without pain	Supine
Brachioradialis	Normal tone in both sides without pain	Supine
Rectus abdominals	Hypotonic in both sides without pain	Supine
Gluteus Maximus	Hypotonic in both sides without pain	Prone
Biceps femoris	Hypotonic in both sides without pain	Prone

Semitendinosus	Hypotonic in both sides without pain	Prone
Semimembranosus	Hypotonic in both sides with no pain	Supine
Rectus femoris	Hypotonic in both sides with no pain	Supine
Vastus medialis	Hypotonic in both sides with no pain	Supine
Vastus lateralis	Hypotonic in both sides with no pain	Supine
Tibialis anterior	Normal tone in both sides without pain	Supine
Gastrocnemius M	Hypotonic in both sides with no pain	Prone
Gastrocnemius L	Hypotonic in both sides with no pain	Prone

Table 12: Initial Muscles Tonicity

3.3.10 Joint Play Examination:

According to **Karel Lewit**

Examined Joint	Result	
	Left	Right
Lisfranc's joints (dorsal \ventral direction)	Free joint	Free joint
Chopart's joint (dorsal \ventral direction)	Free joint	Free joint
Talocrural joint (dorsal \ventral direction)	Restricted joint dorsally	No restriction
Knee joint (medial \ lateral direction)	Free joint in both direction	Free joint in both direction
Patella (all directions)	Restricted laterally	Restricted laterally
Head of fibula (dorsal \ventral direction)	No restriction	No restriction
Interphalangeal joint	Free joint in all direction	Free joint in all direction

Metacarpophalangeal joints	Free joint in all direction	Free joint in all direction
Carpometacarpal joint	Free joint in all direction	Free joint in all direction
Wrist joint	Restricted Palmar shifting	Restricted Palmar shifting
Elbow joint (medial \ lateral direction)	Free joint in all direction	Free joint in all direction
Shoulder joint (ventrodorsal, caudal direction)	Restriction caudal direction	Free joint in all direction
Acromio-clavicular joint (ventrodorsal-craniocaudal)	Free joint	Free joint
Sterno-clavicular joint (ventrodorsal-craniocaudal)	Free joint	Free joint

Table 13: Initial Joint play examination.

3.3.11 Initial Examinations Conclusion:

A 62 years old female patient is suffering from necrotizing autoimmune myopathy (NAM). After the examinations that we made, we found the following findings:

Examinations have been done within 2 days due to patient disease we can say that the patient chief complain is dropped head and marked weakness of neck muscles. Also moderate shortness of pectoralis major and minor. Hypotonic muscular overall the body due necrotizing of the muscles and marked weakness of muscles of upper extremities are noticeable.

Concerning the lower extremities, we can say the weaknesses of muscles are markedly noticeable proximally in flexors and extensors of the knee, hypo-tonicity of

the legs muscles. In general, the patient is able to stand up without assistance for couple of seconds then she needs to support herself for stability, and walk with crutches as well. As it is shown in gait examination we can simply say that lack of stability is seen due to marked weakness of hip adductors.

While examining the joints mobility on both sides we found out that there was restriction in wrist motions in palmar direction (shifting). Also restriction of left shoulder joint in the direction of caudal springing was found. For the knee joint is laterally restricted as well as the patella, in ankle joint (Talucrural joint) is restricted dorsally.

In fact, there is no pain during any motion of both lower and upper extremities, there is only the feeling of exhaustion and being tired after short time exercises or effort is being given. The range of motion is limited in all segments due to weakness of the muscles only not because of restriction or pain, which can be seen between the active and passive motion examinations.

We can say that the patient in a complicated situation where the medications are being changed continuously and the physical therapy is recommended only if the medication works properly. In that case we can work on mainly on strengthening muscles as well as maintaining the good level of activity of the patient. And this is what my rehabilitation plan based on.

3.4 Short & Long Term Plans

3.4.1 Short Term Plan

- Strengthening the weak muscles mainly by active movements against gentle resistance, especially neck flexors and extensors as well as lower extremity for better ambulation.
- Breathing pattern education during exercises by using diaphragmatic breathing.
- Increase the range of motion (ROM) in both upper and lower extremity using methods such as passive stretching.

- Lengthening the shortened muscles especially hip flexors by passive long stretching and Post facilitation stretch according to Janda,
- Relaxation of tight or hypertonic muscles using soft tissue techniques and post Isometric relaxation.
- Reeducation of verticalization and transferring from and to bed.
- Correction of walking stereotype.
- Mobilize restricted joints especially wrist joint in both hands, patella, shoulder joint and ankle joint as well according to Karel Lewit.
- Improve the stability of the patient, awareness of standing and weight distribution and standing with head forward.
- Improve ADL.

3.4.2 Long Term Plan:

- Strengthening overall body muscles exercises by active resisted movements especially neck flexors such as (SCM and scaleni) and hip flexors such (illiopsoas and QF) and biceps & triceps brachii.
- Increase ROM for shoulder joint by flexing, extending, external and internal rotating the joint passively for 10 times each direction. As well as lower extremities for hip joint in direction of flexion and extension.
- Stretching exercises for shortened muscles such as Trapezius, Pectoralis and hip flexors and adductors by techniques such as PSF and passive stretching.
- Relaxation of tight or hypertonic muscles using technique such as PIR and STT.
- Gait training and reeducation, trying with and without crutches.
- Improve and control the transfer progress.

3.5 Therapy Progress: (Day by Day)

Day #1 / Wednesday 1st of February

Objective: the patient doesn't feel any pain during rest, difficulty during transferring from bed to setting and standing next to bed.

Subjective: the patient feels better than the beginning of hospitalization without pain only

during transferring, the patient asked for smaller pillows because the previous pillows were big which makes the shoulder higher and the patient didn't feel well during that, that's why patient complain of

Goals Of Today's Therapy:

- Strengthening the neck flexors and extensors muscles.
- Breathing reeducation and awareness during the exercises.
- Strengthening the upper extremities muscles, handgrip and grasping as well.
- Relaxation of upper trapezius by PIR and STT.
- Increase range of motion of upper extremities.

Implementation:

- Today's therapy was divided into two parts: the first part was focusing on strengthening the neck extensors muscles, mainly was gentle isometric contraction for neck extensors against the pillow while supine.
- I've noticed that the patient is hardly breathing during contraction of any muscles or doing some exercises so I will try to aware the patient of this and next session will be education of breathing.
- The second part was mainly strengthening for upper extremities muscles (biceps and tricep brachii, deltoid and Serratus anterior) by active exercises against soft resistance done by me.
- PIR exercise was done in supine position for both sides to relax the upper trapezius muscles.
- Soft tissue technique by softball was done to both upper Trapezius muscle after the relaxation.
- Active and passive movements were applied to the shoulder joint to increase the range of motion. Isometric contraction with very gentle resistance was applied for biceps and triceps brachii.
- Strengthening of handgrip by pressing on softball.
- The therapy lasted 20 minutes.

Results:

Objective: the patient felt tired during the therapy but she hardly tolerated. Although that we exercised inside her room, it was a bit difficult to manipulate the patient and change positions so I asked if possible to exercise in the rehabilitation room. I will be informed tomorrow-depending patient's condition

Subjective: the patient felt relaxed left shoulder joint after the therapy. On another hand the patient felt spasm in left forearm, decreased sensation in both ankle joints. And she asked to reduce the time in case she felt excessive exhaustion.

Day #2 / Tuesday 2nd of February

Objective: the patient doesn't feel any pain on bed. It seems that she is more tired than yesterday. So, we will exercise in the patient's room for today.

Subjective: the patient spasm in left hip joint, as well as left ankle. She is a bit tired today due to lack of sleep last night. The patient asked to not have hard exercises and no changing of positions if possible.

Goals of today's therapy:

- Prevention of DVT.
- Strengthening the lower extremities muscles.
- Stretching hamstrings and hip adductors, also ankle plantaris.
- Increase ROM of ankle joint motion.

Implementation:

- Today's therapy was focused on lower extremities; we started with simple prevention of thromboembolism in ankle joints, (dorsal and plantar flexion also, circulation). Then we continued with active and passive movements (flexion, abduction and extension in both hip joints), then active and passive movement of knee joint (flexion and extension).
- I continued the session with isometric contraction against gentle resistance in hip joint in the direction of adduction (adductors longus, adductor brevis, adductors

magnus, adductors minimus, gracilis)* as well as knee joint in the direction of extension (Quadriceps femoris muscles).

- After short break, I did passively long stretching for hamstring muscles in supine position, then also passive stretching for long and short adductors for both sides (adductors longus, adductor brevis, adductors magnus, adductors minimus, gracilis), at the end, I moved to ankle joint to stretch the shortened muscles (The gastrocnemius medial head, gastrocnemius lateral head, soleus, plantaris, tibialis posterior).

Results:

Objective: The patient was willing to cooperate even thought she is a bit exhausted today more than yesterday. She tolerated the therapy session, but we had to decrease the duration to 15 minutes twice. There wasn't improvement found in general and the patient status doesn't allow for more examination.

Subjective: The patient felt well in the left ankle joint movements both sides, although the hip joint is still the same.

Day #3 / Friday 3rd of February

Objective: the patient feels excessive pain after biopsy examination.

Subjective: the patient is unable to exercise today due to excessive exhaustion after biopsy examination of both upper and lower extremities.

Goals of today's therapy:

The exercises are cancelled due to patient condition.

Implementation:

Today's therapy was canceled due the patient condition.

Results:

Objective: no physical exercises were applied therefor no results can be found.

Subjective: no physical therapy was applied therefor no results can be found.

Day #4 / Monday 6th of February

Objective: the patient seems well today. She doesn't feel pain.

Subjective: The patient feels better than last week. She feels slight pain in left shoulder, also spasm in left forearm.

Goals of today's therapy:

- Prevention of DVT.
- Reeducation of breathing.
- Mobilization of the shoulder joint.
- Mobilization of wrist joint in both sides.
- Stretching both hamstrings and adductors muscles.
- Strengthening the neck flexors and extensors muscles.
- Strengthening hip muscles (flexors, extensors, adductors and abductors).
- Verticalization and ambulation around the room.

Implementation:

- I started with active ankle joint exercises, by asking the patient to do dorsal and plantar flexion also, circulation.
- Re-educate the patient breathing, by asking the patient to try contracting the diaphragm properly and taking the breath slowly and deeply, I try verbal and tactile technique.
- I exercised the patient's neck while supine by placing patient's head on small medicine ball and as the patient to extend her head against the ball gently, also flexing her head slowly without any resistance at this moment.
- The patient pointed at the beginning of therapy that there is slight feeling of pain in left shoulder anteriorly during the end of motion approximately at 110° of flexion. So I did shoulder traction technique in supine combined with PIR according to Karel Lewit to relax anterior deltoid. Then I did passive movements to the shoulder joint to examine the range of motion.

- Then we continued with active movements without resistance (flexion, extension, abduction and adduction in both hip joints), then active movement of knee joint (flexion and extension). After that I added gentle resistance in the direction of adduction and abduction as the patient is enable to do these movements easier, by isometric contraction against medicine ball in hip joint in the direction of adduction and therapist hands in the direction of abduction, the muscles are targeted (QF, hamstring, biceps femoris, semitendinosus, semimembranosus and adductors muscles*)
- I passively stretched hamstring muscle in both thighs to the 80° then hard barrier appeared.
- We finished the therapy by education of verticalization and walking with crutches around the room almost 2-3 minutes.

Results:

Objective: The patient tolerated the whole therapy session, which lasted almost 45 minutes. There is slight improvement in range of motion in the shoulder joint in the direction of flexion and abduction. Also it is important to mention that there is better and stronger handgrip. For lower extremity, there is slight progress of the stereotype of doing the movements of knee joint as well as range of motion has increased almost 100° in both knee.

Subjective: The patient feels quiet fine after the therapy and she feels her shoulders and ankles in better condition. Also it was good to hear that she can lift her head even though she felt a bit dizziness after standing up otherwise she feels a slight progress in her case. In general she feels much better than last week.

Day #5 / Tuesday 7th of February

Objective: The patient seems fine and nothing in particularly aches.

Subjective: The patient addressed that she was walking with crutches in the morning around the corridor without assistance but she felt dizzy and stopped immediately.

Goals of today's therapy:

- Stretching the shortened hip flexors muscles.

- Reeducation of breathing and diaphragmatic breathing exercises.
- Examine and restore the joint mobility in ankle, wrist and shoulder joint.
- Strengthening neck flexors muscles (longus capitis, longus colli, sternocleidomastoid and anterior scalene muscles).
- Strengthening hip muscles (flexors, extensors, adductors and abductors).
- Strengthening upper limb muscles (deltoid anterior, biceps and triceps brachii, and serratus anterior, Pectoralis major and minor.
- Stretching plantar flexors muscles in ankle joint.
- Walking reeducation in the room and stairs if possible.

Implementation:

- I started today's therapy with mobilization of restricted joints ankle, patella, fibular head and wrist joints.
- The patient is asked to do abdominal breathing by contracting the diaphragm and abdominal muscles deeply and slowly.
- The patient exercised the neck muscles, mainly was gentle isometric contraction for neck extensors against little resistance of medicine ball. Then I added some simple rotations of head using ball under head in supine position associated with visual synkinesis.
- The patient was requested to do active shoulder flexion and abduction, which was improved to almost (flexion to 100° in both shoulder and abduction in right shoulder to 100° and left to 90°). Then I tried to do flexion passively and reached to left 135° and right 150° and abduction passively in the left and reached to 110° and right to 140°)
- Strengthening for upper extremities were applied against resistance mainly biceps, triceps brachii by asking the patient to do flexion and extension of the arm, also Serratus anterior, pectoralis major and minor, by asking the patient to do flexion of the arm and adduction at the level of 90°, for 10 times repetition.
- Then we repeated the exercise of yesterday, with active movements without resistance (flexion, extension, abduction and adduction in both hip joints), then

active movement of knee joint (flexion and extension). After that gentle resistance in the direction of adduction and abduction by isometric contraction against medicine ball in hip joint in the direction of adduction and therapist hands in the direction of abduction, the muscles are being strengthened (QF, hamstring, biceps femoris, semitendinosus, semimembranosus and adductors muscles*).

- I examined the length of hamstring muscles in both thighs and the result is 80°, so I will continue with passive stretching to maintain the physiological length of the muscle.
- I used PSF according to Janda, to stretch gastrocnemius in both lower limbs.
- I examined the joint play and no restriction was found in both lower and upper extremities' joints.
- We finished the therapy by walking with crutches in the room almost 5 minutes, and then she felt tired so, the patient needed to relax and we finish for this day. We couldn't continue walking on stairs for today*.

Results:

Objective: the patient tolerated the therapy session, which lasted 45 minutes. There was slight improvement of range of motion in shoulder and hip joint. No restricted joints were found after mobilization was done. There is still slight shortness of both hamstring and pectoralis major but for the other muscles such as plantar flexors and hip adductors become normal muscle length. The patient is still having some problem of up righting the head position due to weakness of neck flexors.

Subjective: the patient seems satisfied with the results even though she didn't expect that at the beginning.

She feels better in ankle joint and during walking also she doesn't feel any aches in shoulder joint except that just hardening of breathing after exhaustion.

***Day #6 / Wednesday 8th of February**

Objective: The patient went early morning for examination such as biopsy and computer tomography (CT). She finished examination after lunch and she feels tired and not able to exercise today.

Subjective: The patient went for examination and she feels so tired and she needs to eat. So, she isn't able to exercise today.

Goals of today's therapy:

The therapy session had been cancelled due to patient's condition.

Implementation:

Today's therapy was canceled due the patient status.

Results:

Objective: no physical therapy was applied therefor no results can be found.

Subjective: no physical therapy was applied therefor no results can be found.

Day #7 / Thursday 9th of February

Objective: the patient seems fine and nothing aches in particular. As it is scheduled, today is the last day in the rehabilitation period in RÚ, she is leaving afternoon so, we will try to exercise and examine everything possible in case we will have time and the patient condition will allow it.

Subjective: it is the last day of rehabilitation, she seems fine and in better condition, she doesn't complain of pain, from the morning she is feeling well and had breakfast and walked couple of times from her room to dinning room and back. She is ready to exercise today and willing to tolerate it to the end.

Goals of today's therapy:

- Diaphragmatic breathing, actively if possible if the patient is well taught.
- Range of motion examination of ankle joint by doing active plantar and dorsal flexion.
- Strengthening upper limb and examination of strength according to kendall.
- Examination of length in upper trapezius and hamstring muscle.
- Examination of the joint play, which was treated previously.
- Strengthening neck flexors muscles (sternocleidomastoid and anterior scalene muscles).
- Ambulation in the corridor and stairs if possible.

Implementation:

- We started with lower extremities, ankle movements were free and easily done, and there was no restriction in the direction of dorsal and plantar flexion, also increasing range of motion in both direction dorsal and plantar flexion actively done as it will be addressed in the final examination of ROM.
- Then I strengthened neck flexors muscles (sternocleidomastoid and anterior scalene muscles)., mainly was gentle isometric contraction for neck flexors against slight resistance of therapist fingers, then rotation of head using ball under head in supine position associated with visual synkinesis.
- Then we repeated the exercise of last week, with active movements without resistance (flexion, extension, abduction and adduction in both hip joints), then active movement of knee joint (flexion and extension). After that gentle resistance in the direction of adduction and abduction by isometric contraction against medicine ball in hip joint in the direction of adduction and therapist hands in the direction of abduction, at this moment there was no big improvement of overall strength but the patient it is quiet clear that adductors muscles are contracting stronger comparing with abductors. There is good progression of ROM in knee as well as hip motions; it will be shown in the lower extremities goniometry.
- Stretching of shortened pectoralis minor muscle in both shoulders. Also the hamstring muscle in both sides by passive stretching. At the final I stretched the upper trapezius in both sides.
- Patient stood up independently and start walking with crutches around the room almost 6 minutes, with still difficulties of up righting the head at the beginning then the patient started to feel tired.
- I asked the patient to show pattern of breathing during all the exercises, the patient showed good progressive of co-activation of diaphragm and abdominal muscles even though it is quiet difficult and tiring.
- Today's therapy was intensive and lasted 60 minutes.

Results:

Objective: The patient is still having problem of up righting the head position due to weakness of neck flexors. There is still slight shortness of both pectoralis minor. It is

important to say that there is still significant weakness of muscles over all but there is slight improvement of strength and range of motion in general.

Subjective: the patient feels much better in general. Handgrip and strength of upper extremities have improved. She is going to continue rehabilitation in Pardubice neurology department because it is next to her living home.

3.6 Final Kinesiological Examinations:

3.6.1 Postural Examination-Aspection-Plumb Line Test:

The patient is able to stand up next to bed for 1-2 minutes then she will be tired. She has difficulty of upright the head position.

Back View	
The base of support	Normal
Shape of the heels	Internal rotated
Position of the ankle joints	Valgus
The calf muscles	Hypo-tone symmetrically
Position of the knee joints	Valgus Knees
Position of pelvis	Elevation of right side
Paravertebral muscles	Hypo-tone
Curvature of the spine	Physiological curvature.

Position of the scapula	Elevation of left scapula
Position of the shoulder girdle	Elevation of left shoulder
Position of head	Forward dropped.

Table 14: Final Aspection of back view

Side View	
Position of the ankle joints	Medially pressed (valgus)
Position of the knee joints	Left knee looks downward than right.
Thigh muscles	Hypo-tone both sides
Curvature of the Spine	Physiological curvature in Thoracic spine and flat Lumbar, and lordosis curvature in cervical spine.
Position of shoulder joint	Elevation of left side
Position of the head	Protracted, dropped forward

Table 15: Final Aspection of side view

Front View	
Weight distribution	Left tendency
Position of the knee joints	Valgus
Shape of the thigh muscles	Hypo-tone both sides
Position of the pelvis	Elevation of right side
Position of the shoulder girdle	Elevation of left shoulder
Position of the head	Forward dropped

Table 16: Final Aspection of Front view

3.6.2 Goniometry – SFTR-:

Hip Joint				
	Active		Passive	
	Left	Right	Left	Right
S	10° - 0 - 110°	10° - 0 - 110°	10° - 0 - 120°	10° - 0 - 140°
F	50° - 0 - 10°	50° - 0 - 10°	60° - 0 - 30°	60° - 0 - 30°
R	50° - 0 - 40°	50° - 0 - 40°	60° - 0 - 45°	60° - 0 - 50°
Knee Joint				
S	0° - 0 - 90°	0° - 0 - 90°	0° - 0 - 120°	0° - 0 - 140°
Ankle Joint				
S	20° - 0 - 40°	30° - 0 - 40°	35° - 0 - 50°	35° - 0 - 50°
R	20° - 0 - 30°	30° - 0 - 35°	30° - 0 - 40°	30° - 0 - 50°

Table 17: Final Goniometry of lower extremities (degree).

Shoulder Joint				
	Active		Passive	
	Left	Right	Left	Right
S	30° - 0 - 110°	30° - 0 - 120°	50° - 0 - 150°	60° - 0 - 160°
F	90° - 0 - 0°	110° - 0 - 0°	160° - 0 - 0°	160° - 0 - 0°
T	90° - 0 - 15°	90° - 0 - 30°	120° - 0 - 30°	120° - 0 - 30°
R	75° - 0 - 65°	75° - 0 - 65°	80° - 0 - 70°	80° - 0 - 70°
Elbow Joint				
S	0° - 0 - 120°	0° - 0 - 120°	0° - 0 - 150°	0° - 0 - 150°
Wrist Joint				

S	70° - 0 - 70°	80° - 0 - 80°	85° - 0 - 90°	90° - 0 - 90°
F	15° - 0 - 20°	20° - 0 - 30°	20° - 0 - 35°	20° - 0 - 35°

Table 18: Final Goniometry of upper extremities (degree)

3.6.3 Gait Analysis:

The patient is able to walk around the room with crutches. Drop head during walking with no rotation of pelvis. The patient has duck walk due to weak hip muscles.

Base of support	Normal
Walking rhythm	Non Periodic
Walking speed	Slow
Stride length (short steps/long steps)	Normal length
Movement of the foot	Heel strike -flat foot- toes off
Axial position of the lower limb	Flat Foot (both)
Position and movements of the pelvis	Swinging normally
Movement of center of gravity (COG)	Not stable
Position and movements of the trunk	No rotation due to crutches
Activity of back and abdomen muscles	Slightly
Position and movements of the head	It is difficult of up righting the head position during walking
Movements of the upper extremity	Synchronicity.

Table 19: Final Gait analysis.

3.6.4 Specific Testing

- **Romberg Test** (I-III): the patient was able to do the test I- correctly without losing her balance but she lost her balance while doing Romberg II-III.
  Romberg I – Negative
  Romberg II – Positive
  Romberg III – Positive

- **Modification of Standing:**

Standing on tiptoes: Positive.

Standing on heels: Positive.

3.6.5 Anthropometric Measurement (length, circumferences)

Upper Extremities	Left	Right
Length of upper limbs	65 cm	66 cm
Length of humerus	25 cm	25 cm
Length of forearm	24 cm	24 cm
Length of hands	17 cm	17 cm
Circumference of upper limb	38 cm	39 cm
Circumference of forearm	33 cm	33 cm

Table 20: Final Anthropometric measurement (upper extremities)(cm)

Lower Extremities	Left	Right
Anatomical length	97 cm	97 cm
Functional length	85 cm	85 cm
Length of thigh	42 cm	42 cm
Length of middle leg	32 cm	32 cm
Circumference of thigh	72 cm	73 cm
Circumference of knee	60 cm	60 cm
Circumference of calf	58 cm	58 cm
Circumference of ankle	45 cm	45 cm
Circumference of foot	21 cm	21 cm

Table 21: Final Anthropometric measurement (lower extremities) (cm)

3.6.6 Spine Distances:

Test	Result

Thomayer	+12 cm
Stibor	7 cm
Shobar	4 cm
Forest Fleche	7 cm
Lateral flexion	24 cm symmetrical

Table 22: Spine distances (cm)

3.6.7 Muscle Length Test:

Muscles	JANDA		KENDAL		Result
	L	R	L	R	
Plantar flexors	-	-			No shortness
Hip flexors					Marked shortness of one and two joints of hip flexors
Hip adductors	-	-			No shortness of both hip
Hamstring			80°		Moderate shortness in both hamstring
Paravertebral Muscles	2				Marked shortness of paravertebral muscles
Pectoralis major- Upper Lower			90° 135°		Normal length of both upper Pectoralis Normal length of both lower Pectoralis
Pectoralis minor					Moderate shortness of both pectoralis minor
Levator scapula	1	1			Moderate shortness Levator scapulae
Trapezius (cranial)	-	-			No shortness was found
Sternocleidomastoid	1	1			Moderate shortness

Table 23: Final Muscles length test.

3.6.8 Muscle Strength Test:

According to Kendall.

Tested Muscle	Left	Right
Soleus	3	3
Gastrocnemius	3	3
Palntaris	3	3
hamistring	+3	+3
Biceps femoris	+3	+3
Hip flexors	3	+3
Tensor fascie latae	-3	-3
Gluteus minimus	-3	-3
Gluteus medius	-3	-3
Gluteus maximus	3	3
Hip Adductors	+3	4
Neck flexors	2	2
Neck flexion with rotation	2	2
Neck extensor	2	2
Deltoid Medially	+3	+3
Anterior deltoid	+3	+3
Posterior deltoid	3	3
Teres major	3	3
Pectoralis major, upper part	3	3

Pectoralis major, lower part	3	3
Pectoralis minor	3	3
Biceps brachii	4	4
Triceps brachii	4	4
Latissimus dorsi	3	3
Infraspinatus	3	3
Levator scapulae	3	3
Trapezius (cranially)	3	3
Rectus Abdominis	2	

Table 24: Final Muscles Strength Test.

3.6.9 Muscle Tonicity:

Tested Muscles	Palpation	Position
Levator scapula	Hypotonic in right side with no pain	Supine
Trapezius	Hypotonic in right side with no pain	Prone
Sternocleidomastoid	Hypotonic in both sides with no pain	Prone
Supraspinatus	Hypotonic in both sides with no pain	Prone
Infraspinatus	Hypotonic in both sides with no pain	Prone
Pectoralis major	Normal tone in both sides without pain	Prone
Pectoralis minor	Normal tone in both sides without pain	Prone
Latissimus dorsi	Hypotonic in both sides with no pain	Prone
Deltoid	Normal tone in both sides without pain	Prone

Biceps brachii	Normal tone in both sides without pain	Supine
Triceps brachii	Normal tone in both sides without pain	Supine
Brachioradialis	Normal tone in both sides without pain	Supine
Rectus abdominals	Hypotonic in both sides with no pain	Supine
Gluteus Maximus	Hypotonic in both sides with no pain	Prone
Biceps femoris	Hypotonic in both sides with no pain	Prone
Semitendinosus	Hypotonic in both sides with no pain	Prone
Semimembranosus	Hypotonic in both sides with no pain	Supine
Rectus femoris	Hypotonic in both sides with no pain	Supine
Vastus medialis	Hypotonic in both sides with no pain	Supine
Vastus lateralis	Hypotonic in both sides with no pain	Supine
Tibialis anterior	Normal tone in both sides without pain	Supine
Gastrocnemius M	Hypotonic in both sides with no pain	Prone
Gastrocnemius L	Hypotonic in both sides with no pain	Prone

Table 25: Final Muscles tone palpation.

3.6.10 Joint Play Examination:

According to Karel Lewit

Examined Joint	Result	
	Left	Right
Lisfranc's joints (dorsal \ventral direction)	Free joint	Free joint
Chopart's joint (dorsal \ventral direction)	Free joint	Free joint

Talocrural joint (dorsal \ventral direction)	Free joint	Free joint
Knee joint (medial \ lateral direction)	Free joint in all direction	Free joint in all direction
Patella (all directions)	Free joint	Free joint
Head of fibula (dorsal \ventral direction)	Free joint	Free joint
Interphalangeal joint	Free joint in all direction	Free joint in all direction
Metacarpophalangeal joints	Free joint in all direction	Free joint in all direction
Carpometacarpal joint	Free joint in all direction	Free joint in all direction
Wrist joint	Free joint	Free joint
Elbow joint (medial \ lateral direction)	Free joint in all direction	Free joint in all direction
Shoulder joint (ventrodorsal, caudal direction)	Free joint in all direction	Free joint in all direction
Acromio-clavicular joint (ventrodorsal-craniocaudal)	Free joint	Free joint
Sterno-clavicular joint (ventrodorsal-craniocaudal)	Free joint	Free joint

Table 26: Final Joint play examination.

3.7 Evaluation of the Effect of the Therapy:

Before the patient leaves the institution of rheumatoid I've done general examination and evaluation for the patient status at the moment, it is so important to

mention that the patient's diagnose is autoimmune which leads to rapid exhaustion after any kind of therapy or little effort of changing position or even moving head. In general there was no significant improvement of correction of dropped head after standing up but the patient now is able to up righting the head after couple of attempt unlike at the beginning of rehabilitation. After doing mobilization to most of joints regardless the spine due to difficulty of changing the position, the joints are free and no restriction was found which led to find out increased range of motion passively and actively. For the matter of muscles strength, due to the patient disease we can't expect much improvement even though we can see slight progress in hand functions especially biceps and triceps brahii, as well as hip extensors gluteus maximus and hamstring of both legs.

The patient clarified that she feels much better in the shoulder joint and the transferring become faster process and it takes less time than used to be. It is important to mention that all practical session and therapy procedures were done on patient's bed due to patient request so it there was some difficulties regarding some therapeutic procedures and techniques, which demand changing positions and adjustable beds. The patient is well trained to self-reliable in transferring and ambulation and she doesn't need any assistance with that at this moment. Even though there is a still difficulty in up righting the head after standing up but the patient is able to achieve.

4 Conclusions:

I would like to emphasize that there was noticeable progression of the patient's status, but since the patient is autoimmune case, the condition of patient might worsened within a week or days due to intervention of medication.

I could possibly say that there are some good improvements regarding the range of motion of both upper and lower extremities, joints mobility and breathing stereotype. But on the other hand, there was no much progression when are talking about muscles tonicity and strength,

At the end I can say that the benefits that I had from this period of time which I spent with patients at **Revmatologický ústav.** It was amazing experience because I really found out how a professional physiotherapist would treat the patient and how would

communicate with them in a professional way and to experience how the full day of my career will be like in the future.

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6 Supplements:

6.1 List of Figures:

6.2 List of Tables:

6.3 List of abbreviations:

NAM: Necrotizing Autoimmune Myopathy.

HMGCR: 3-Hydroxy-3-Methylglutaryl-CoA Reductase.

DM: Dermatomyositis.

PM: Polymyositis.

JDM: Juvenile Dermatomyositis.

sIBM: Sporadic Inclusion-Body Myopathy

CK: Creatine kinase.

IIM: Idiopathic Inflammatory Myopathies.

SRP: Signal Recognition Particle.

AIM: Auto Immune Myopathies.

HIV: Human Immudefency Virus.

SINAM: Statin Induced Necrotizing Autoimmune Myopathy.

MHC-I: Major Histocompatibility Complex-I.

CTD: Connective Tissue Disease.

EMG: Electromyography.

MRI: Magnetic Resonance Imaging.

PET: Positron Emission Tomography.

SGOT: Serum Glutamic Oxaloacetic Transaminase.

SGPT: Serum Glutamic Pyruvic Transaminase.

MTX: Methotrexate.

IVIg: Intravenous Immunoglobulin.

STT: Soft Tissue Technique.

SCM: Sternocleidomastoid

ADL: Activity of Daily Living.

ROM: Range Of Motion.

PIR: Post-Isometric Relaxation.

PSF: Post Facilitation Stretch.