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Knowledge, Attitude, Lifestyle Practices, and Quality of Life in Sporadic Lymphangioleiomyomatosis Patients

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by

Shahpar Vafamand

MS, George Mason University, 1994

BS, Eastern Kentucky University, 1983

Dissertation Submitted in Partial Fulfillment

of the Requirements for the Degree of

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Abstract

Lymphangioleiomyomatosis (LAM) is a rare lung disease recognized by abnormal growth of smooth muscle cells proliferating in lungs parenchyma, developing benign tumors, migrating to the other organs, and ultimately leading to respiratory failure and death. Despite existing literature mainly on clinical aspects of LAM, there is a gap of literature in regards to the knowledge, attitude, and lifestyle practices (KAPs) of LAM patients and their effects on their quality of life. The purpose of this quantitative study was to investigate the KAPs of the sporadic LAM patients as measured by the Bristol Chronic Obstructive Pulmonary Disease Knowledge Questionnaire, Beliefs and Behavior Questionnaire, Determinants of Lifestyle Behavior Questionnaire; these KAPs were then analyzed for their relationship to quality of life reports as measured by the St George's Quality of Life Questionnaire. Transtheoretical model (TTM) was used to describe the relationship among the variables. The data were collected through online survey questionnaires from 143 sporadic LAM patients registered at the LAM Foundation. Pearson's correlations and linear regression were used to analyze the data. The results of the analysis showed that there was a significant positive relationship between attitude, lifestyle practices, and quality of life and a negative relationship between knowledge and quality of life. The outcome achieved by this study and its implication on social change identifies the need to initiate more study-specific KAPs within LAM populations, including individuals with tuberous sclerosis complex LAM. The results could also encourage the LAM community as well as other stakeholders to implement programs, workshops, and interventions that could promote and enhance quality of life.

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Dedication

I dedicate my entire dissertation to my mom and dad. To my father who had already painted this picture when I was a child. Thanks for the discipline and courage you instilled in me to never bend no matter how heavy my load of challenges got. To my Mother who I can't find any words to describe how deeply I love and admire. Your sacrifices and endless love has made me the person that I am today. You have always exemplified the person who I wanted to be. Your examples have taught me to be strong and to never give up working hard for the things I aspire to achieve. I love you with every beat in my heart, and I dedicate to you one of the most valuable accomplishments in my life.

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Table of Contents

Chapter 1: Introduction to the Study

Lymphangioleiomyomatosis (LAM) is a rare cystic, multisystem lung disease

that almost exclusively affects women of childbearing age (King, 2010). LAM is

characterized by progressive cystic destruction of the lung and lymphatic and abdominal

tumors (Ryu et al., 2006). LAM is recognized by abnormal growth of smooth muscle

cells proliferating in lungs parenchyma, developing benign tumors known as

angiomyolipomas (AMLs) in kidneys, benign abdominal tumors known as

lymphangiomyolipomas, migration to the other organs such as liver and brain ultimately

leading to respiratory failure and death (Johnson, 2006). The most common symptoms

are shortness of breath or dyspnea which is present in 42% of patients and eventually

universal as disease moves further along (Almoosa, Ryu, & Medez, 2006). Another

complication of LAM is pneumothorax which occurs in over 70% of the women with

LAM at some point in their disease course, and with chances of reoccurrence among two

thirds of these target population (Johnson, Lazor, & Cordier, 2002). LAM is found

sporadically in patients with no genetic abnormality and in over 30% of women with

tuberous sclerosis complex (TSC) which is an inherited autosomal dominant tumor

suppressor syndrome (McCormack, 2008) characterized by hamartomas, seizures, and

mental retardation (Avila, Dwyer, Rabel, & Moss, 2007). Although there are some

similarities between sporadic LAM and TSC LAM, the differences are substantial

between the two groups of patients (LAM Foundation, 2013). According to Avila et al.

(2007), the severity of lung disease is much more extensive in patients with sporadic

LAM than TSC LAM, (60% vs. 37%) respectively. Patients with TSC LAM have more

Renal AMLs (93% vs. 32%), hepatic AMLs (33% vs. 2%), and higher incidences of noncalcified pulmonary nodules (12% vs. 1%; Avila et al., 2007). It was not within the scope of this research to examine both TSC and sporadic LAM patient's knowledge, attitude, and lifestyle practices (KAPs), therefore the intention of this research was to focus on the population with the sporadic LAM which also tends to be 60% to70% of LAM population (LAM Foundation, 2013).

LAM is often misdiagnosed due to the rarity of this disease and lack of awareness among the health care community to recognize the signs and symptoms of this disease. It is very common for the health care providers to miss proper diagnosis of women with LAM due to the fact that its presentation of symptoms resembles other respiratory diseases such as asthma or emphysema (LAM Foundation, 2013). Although LAM was first introduced to the medical literature by Von Stossel in 1937, it was not until 1997 that a national LAM registry was established by the National Heart, Lung, and Blood Institute (NHLBI) in order to seek a more accurate demographic, clinical, physiologic, and radiologic characteristic of women with LAM (Ryu et al., 2006). There is no cure for LAM and the treatment for women with this debilitating disease is mainly supportive (Cottin, Mortex & Archer, 2011). There has been tremendous effort by the LAM Foundation, LAM alliance, and other organizations to promote awareness and education not only for the health care communities but also patients and their families. Despite all their efforts and the existing literature, there were no prior studies that have addressed the importance of knowledge about LAM, patients' attitudes about LAM, or lifestyle practices impacts' on the quality of life of women with LAM. Knowledge can be

essential not only for the health care practitioners but also for the patients to understand how well they could manage living with LAM. Improved attitude and healthier lifestyle practices could also allow these targeted populations to improve the overall quality of their lives. The objective for this study was to examine the effects of KAPs of the LAM patients on their quality of life. The goal of this study was to encourage a social change through possible desired outcome to enhance the quality of life for women with LAM.

Background

The existing literature was predominately in regards to the clinical aspects of LAM; thereby leaving noticeable gap in the area of the patients' KAPs and their effects on their quality of life. Von Stossel recognized LAM in 1937 and described it as a multisystem disease proliferated through abnormal growth of smooth muscle cells infiltrating through lymphatic and lungs (Ryu et al., 2006). Since then, there has been tremendous improvement and efforts in understanding LAM and disease description through obtained information from the LAM Foundation, which, according to Harknett et al. (2011), holds the largest patient profile registry as well as the National Heart and Blood Institute of Health (NHLBI). The information collected and shared with clinicians involves rate of lung decline; lung transplantation, as well as comorbidities related to LAM (Cottin, et al., 2011). Throughout the years since first cases of LAM appeared in 1937, milestones for LAM research have revolved (a) around comprehensive identification of LAM patients, (b) identification of the *TSC1* and *TSC2* genes coding for the tuberin and hamartin proteins respectively, (c) recognizing the *TSC2* mutations in Renal and pulmonary LAM cells, (d) NHLBI LAM registry, (e) discovery of the

mutations of the *TSC2* gene, (f)observation of LAM cells regenerating in the donor lung
after transplantation (Karbowniczek et al., 2003), and (g) the sirolimus trial in 2011
which was promising in improving and stabilizing lung function as well as reducing
pleural effusions, and AMLs (Taveira-DaSilva, Hathaway, Stylianou & Moss, 2011).

     For the purpose of this investigation, I researched the effects of KAPs and quality
of life in patients with similar conditions which are described more in detail in Chapter 2
of this research. Researchers have emphasized the importance of knowledge and
education in understanding one's health condition, its relation to the patient's attitude,
and quality of patient's life has been emphasized in other chronic diseases as well. For
instance, Boot et al., (2005) reported that their results showed association between high
knowledge about how to manage the disease; adequate adaptation which meant becoming
aware and accepting of their chronic illness; and limitations to their daily functions. He
also elaborated that gaining knowledge about one's disease would promote perceived
control over health that would most likely affect behavior change. Lua and Neni (2011)
studied awareness, knowledge, attitude and their importance in quality of life of people
with epilepsy; the authors concluded that incorrect knowledge and awareness caused
more suffering than the disease itself. They claimed that knowledge and awareness along
with positive attitude had a noticeable relation to the patient's quality of life; and as result
of their findings Lua and Neni (2011), concluded that it is essential to initiate educational
initiatives to continuously improve on awareness, knowledge, and attitude levels to
impact quality of life.

This study was needed because it was an important step in recognizing the gap in the literature in regards to the role of KAPs and LAM patients quality of life. The aim of this research was to possibly encourage and promote a social change within the LAM community. The research findings could motivate the stakeholders and health care professionals to embrace and initiate interventions that would promote knowledge, attitude, and healthy life style practices. These initiatives could improve quality of life for LAM patients who are faced with physical and emotional challenges for their entire remaining lives. (LAM Foundation, 2013)

Problem Statement

Despite tremendous focus in studying the clinical aspect of LAM and available literature on etiology of LAM, diagnostic criteria, and available treatment for pulmonary and abdominal complications in women with LAM, quality of life within LAM population has been an ongoing challenge and lack of research in this area in particular has created a noticeable gap in literature.

The rarity of LAM has created many challenges for the practitioners to recognize the signs and symptoms of this disease. The similarities with LAM and other respiratory diseases such as asthma; bronchitis, or emphysema have delayed the proper diagnosis of women ultimately causing concerns; confusion, and affecting quality of life with this debilitating disease (Zhang & Travis, 2010). It would be important to initiate more studies and focus in the role of KAPs and how they relate to the quality of life in LAM population.

Purpose of the Study

The purpose of this quantitative study was to examine the KAPs of the study population and their relation to quality of life among women living with sporadic LAM. This study utilized the quantitative methodology incorporating the survey research strategy in order to collect data. The population that participated in this study was women who had been diagnosed with sporadic LAM. I collected new data and was able to determine the knowledge of these targeted women about LAM; the attitude they hold towards the quality of life; and the life style practices they have living with LAM

The outcome achieved by this study and its implication on social change could encourage researchers to initiate more studies on KAPs within the LAM population, in particularly future research on the KAPs of the TSC LAM population as the focus of the present study was women with sporadic LAM. The results could also motivate the stakeholders to promote initiatives that would improve KAPs that could enhance and create much needed hope and encouragement to possibly enhanced quality of life.

Research Questions and Hypotheses

Research Question 1: To what extent does level of knowledge about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 1: There is an influence of the knowledge about LAM in women living with LAM on their quality of life.

Null Hypothesis 1: There is no influence of the knowledge about LAM in women living with LAM on their quality of life.

Research Question 2: To what extent does the attitude about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 2: There is an influence of the attitude about LAM in women living with LAM   on their quality of life.

Null Hypothesis 2: There is no influence of the attitude about LAM in women living with LAM on their quality of life.

Research Question 3: To what extent do the life style practices of women living with LAM influence their quality of life?

Alternative Hypothesis 3: There is an influence of life style practices of women living with LAM on their quality of life.

Null Hypothesis 3: There is no influence of life style practices of women living with LAM on their quality of life.

Theoretical Framework

The theoretical framework for this research was the transtheoretical model (TTM), or stages of change. Adaptation of such a theory was ideal for this study because it focuses on individuals adopting healthy behaviors and letting go of the unhealthy behaviors. According to Prochaska and Velicer (1997), through the six stages of change such as precontemplation, contemplation, preparation, action, maintenance, and termination individuals would prepare and accept the changes knowingly and willingly. The TTM assumes that individuals do not change behaviors quickly: rather, change happens in a continual pattern and the possibility of relapse to the previous step is quite possible. The TTM allows for interventions to be designed specifically for targeted

populations' need and desire to change behavior. In the case of this research, the TTM and the stages of change would stimulate the desire in LAM patients to become knowledgeable in all aspects of LAM and empowered through gained knowledge to change behavior and adopt healthier lifestyle practices. Intervention strategies could motivate and assist LAM patients to go through each stage starting with precontemplation. At this stage women are most likely confused and do not even think changing behavior could be beneficial; yet once women start accepting the need for change in behavior in the contemplation stage more pros than cons are considered (Prochaska & Velicer, 1997). The stage of determination, or action, is going through the change and embracing the decision to adopt needed behavior towards practices that could ultimately alter the LAM patient's quality of life. Going through previous stages of change will become leading steps to action and maintenance stages of TTM model. The stages of change model related to this study because it could encourage women to increase their awareness and understanding of LAM and become motivated to change or adopt a behavior and lifestyle practices that would be conducive in obtaining or altering their quality of life.

Nature of the Study

This research study utilized the quantitative study design in order to examine the KAPs of the LAM population participating in the study; and their effect on the quality of life. The study population was women who had been diagnosed with sporadic LAM. The survey research design was incorporated in order to collect data from the study participants. The research questions in this quantitative study evaluated the knowledge of

the patients in regards to LAM, their attitude and their lifestyle practices, and what effect those had on their quality of life. Through my review of related literature, I was able to identify the gap in literature in regards to KAPs of the sporadic LAM patients and quality of life which signified the need to research such variables and their effects on the outcome variable. I provide more details in regards to the data collection and data analysis in Chapter 3 of this research study.

Definitions of Terms

Angiomyolipomas (AMLs): Tumors containing fat, smooth muscle, and blood vessels that can infiltrate the kidney and cause renal failure (McCormack et al., 2008). AMLs occur in approximately 80% of the women with LAM (Avila et al., 2000).

Chronic obstructive pulmonary disease (COPD): Chronic disease that involves progressive airflow limitation representing symptoms of dysepnea, cough, and sputum production (Hernandez, Balter, Bourbeau, & Hodder, 2009).

Chylous effusions: Obstruction of lymphatic vessels by the excess muscle growth that can leak fluid in to the chest cavity at time the fluid can contain fat and milky white appearance (NHLBI, 2011).

Lymphangioleiomyomatosis (LAM): Progressive, proliferative, and infiltrating smooth muscle cell that will lead to the cystic destruction of the lung parenchyma; obstruction of the airways, blood vessels, lymphatics, and loss of pulmonary function (Zhang & Travis, 2010).

Lymphangioleiomyomas: Proliferation of smooth muscle cells in lymph vessels that can produce cystic masses and it occurs in 20% of LAM population (Pallisa et al., 2002).

Pneumothorax: Frequent complication of LAM which is caused by air leak to the chest cavity. It occurs in almost 50% of women and 70% chances of recurrence (Young et al. 2006).

Sporadic LAM: Lymphangioleiomyomatosis that is present in patients with no genetic evidence and it is mostly caused by mutations in the TSC2 genes (Taveira-DaSilva et al., 2011). This form of LAM affects approximately 1 in a 1,000000 women in the world (Cho et al., 2010).

Tuberous sclerosis complex (TSC LAM): Autosomal dominant disorder characterized by the development of hamartomatous tumors in brain, kidney, skin, retina, heart, and lungs which affects 30-40% of women with LAM caused by germline mutations of either the TSC1 or TSC2genes (Finlay, 2004; Seibert et al. 2011).

<div align="center">**Assumptions**</div>

It was assumed that the participants in the study accurately responded to the survey questionnaires and sufficiently answered all the questions asked. The other assumption in this study was that the participants' willingness to participate and respond did not cause any bias in the study. It was also assumed that the questions in the survey were appropriate means for collecting data in order to measure and analyze the variables. The analysis of the data was based on the answers given by the participants and it was

assumed that these respondents were completely cognizant of their answers and did not knowingly exaggerate to make their points.

Scope and Delimitation

This study research was limited to the sporadic LAM population; therefore the results are applied to those women who were diagnosed with sporadic LAM. The findings limit generalizing the results of the study to the TSC LAM patients.

The delimitation imposed on this study was that the women who participated in the study were from the largest registry of LAM patients up to date consisting of 1,775 patients in which 84% consisted of sporadic LAM patients (LAM Foundation, 2013). The readily accessible LAM population allowed for the ease of sampling process, although this delimitation restricted the populations to which the outcome of this study could be generalized. The findings of this study were limited to the sporadic LAM participants.

Limitations

One of the limitations in this study was the fact that study participants were from only one patient registry and this limited the generalizability of the results to the rest of the LAM population. The other limitation was the fact that the study population was limited to only sporadic LAM patients and not TSC LAM patients, which would restrict the findings to only serve the sporadic LAM patients and not TSC LAM patients. A further limitation was that LAM registry population could have been more educated in regards to LAM than other LAM patients elsewhere, and this could have been be a limiting factor and cause for bias. The fact that I am a LAM patient myself could also be another reason for bias; however, this research was solely done based in an objective and

scholarly manner. Additionally, because the study questionnaires were not specifically designed for LAM it could have been a cause for bias and limitation to this study.

Significance

This study is significant because it addresses the gap in the literature in regards to the importance of patient's knowledge and attitude about LAM; healthy lifestyle practices, and their relationship to quality of life. LAM patients are living with a disease that is rare, prograssive, and most often fatal (King, 2010). Treatment of LAM is mostly supportive with supplementation of oxygen if needed and treatment of complications such as pnemothorax (Cottin et al., 2011). The possabilities of improving quality of life through gained knowledge, positive attitude, and healthy lifestyle practice could promise a positive social change for the LAM community. The findings of this study could encourage LAM communities as well as other stake holders to implement programs, workshops, and interventions that could promote such goals. Through this study's results, these targeted women could reap the benefits of improved quality of life through KAPs that can be conducive to achieving such desired outcomes.

Summary

LAM is a rare multisystem lung disease occurring almost exclusively in women; affecting not only lung but also lymph nodes and abdominal tumors (Harari, 2011). Although there has been great improvement in organizations and research in clinical and etiology of LAM, there are no studies looking in to the KAPs and their effects on quality of life in LAM patients. Although there was no history of any research in regards to the importance of this matter within the LAM community, there are literatures in regards to

the role that KAPs play in the quality of life in patients with the similar chronic diseases.

Literature research is covered in great detail in Chapter 2. The methodology section

would be covered in Chapter 3, which will address the data analysis and measurement.

This chapter will also discuss the research design, sample population, and

instrumentation utilized to collect data.

Chapter 2: Literature Review

Introduction

This chapter includes the current literature on LAM and its etiology, types of LAM, clinical presentations, pulmonary physiology, diagnosis, and available treatment. Due to the rarity of LAM disease; studies within the last decades have been primarily focused on the clinical aspect of LAM and not on KAPs of LAM patients and their relation to their quality of life; therefore, I have provided existing literature on the importance of KAPs and health- related quality of life in other chronic diseases. This literature review will establishes the need for research concerning the association between the KAPs of women with LAM and their importance in their quality of life. The theoretical framework for this dissertation relied on the TTM or stages of change. Prochaska and Velicer (1997) stated that the importance of this theory is rooted in the six stages of change: precontemplation, contemplation, preparation, action, maintenance, and termination. The theoretical model suited this research because it showed the process in which patients change behavior through stages. Although not everyone would adhere to the particular stage at all times nor change behavior at the same level at all times; this framework lays out a path to look in to the effects of change from the precontemplation to the maintenance stage adopting behaviors conducive to quality of life.

In my efforts to collect scientific literature, I obtained journal articles from a variety of databases and extensive searches through medical and health science databases such as MEDLINE, CINAHL, Science Direct, SAGE, Orphanet, PubMed, Google Advanced Search, and Walden library. The key terms used for this literature search were

Lymphangioleiomyomatosis, sporadic LAM, tuberous sclerosis complex (TSC) LAM,

cystic lung disease, pneumothorax, angiomyolipomas, Chylous effusions, knowledge,

awareness, attitudes, health-related quality of life, healthy life-styles, risk behaviors,

KAPs , chronic disease, lung disease, chronic obstructive lung disease, pulmonary

disease, patient education, and *attitude.*

LAM

LAM is a rare, progressive, multisystem, and cystic lung disease that was first

introduced to the medical literature by Von Stossel in 1937. A national LAM registry was

established by the NHLBI in 1997 in order to seek a more accurate demographic, clinical,

physiologic, and radiologic characteristic of women with LAM (Ryu et al., 2006). LAM

is found almost exclusively in women of childbearing age with a mean onset occurring

between the ages of 34 and 39 (Ryu et al., 2006). Although presentation of this disease is

less seen in postmenopausal women, the reported case series by the NHLBI registry

showed 40% of 230 women were postmenopausal (Ryu et al., 2006). In a cross-sectional

study of 507 LAM patients, Cohen, Pollock-BarZiv and Johnson (2005) reported that the

mean age of women with diagnosis of LAM was 42.7 years in comparison to reported

mean age of 35.8 and 30.2 years one and two decades ago respectively. Cohen et al. 2005

concluded that older women were being diagnosed with LAM, therefore LAM should not

be thought of as a disease of childbearing age women only and needs to be broadened to

include the middle- aged women as well.

LAM is destructive to the lungs; causing lymphatic abnormalities and abdominal

tumors known as AMLs (Noonan & Lou, 2010). LAM is characterized through abnormal

growth of smooth muscle cells which has the ability to grow within the pulmonary airway, lymphatic, blood vessels and gradually lead in to respiratory failure and death (Zhang & Travis, 2010). The disease can occur either sporadically, which affects 1 in 400,000 women (Cordier & Lazor, 2005) or in people with TSC-LAM with 30% to 40% prevalence among adult females (Seibert et al., 2011). Sporadic LAM occurs for unknown reasons and almost never in men (NHLBI, 2011). Patients with sporadic-LAM present with almost the same extra-pulmonary features as TSC-LAM such as renal AMLs and abdominal lymphangiomyomas; however they do not have the symptoms on their skin, central nervous system or the eye manifestations (Ryu et al., 2006). The TSC-LAM is an autosomal dominant disorder that is recognized by the hamartomatous tumors in several organs such as brain, retina, skin, heart, and lung (Seibert et al., 2011). TSC-LAM is caused by mutations in *TSC1* or *TSC2* genes which encode the proteins hamartin and tuberin respectively. The mutations in either one of these genes are sufficient for the TSC to be present; however the mutation in *TSC2* genes is strongly associated with sporadic LAM. These two proteins are responsible for inhabiting the growth factor stimulation of mammalian target of rapamycin (mTOR) and once one of these genes are mutated the signaling fashion is disturbed and would encourage the abnormal smooth muscle cells growth (Johnson et al., 2002; Ryu et al., 2006).

Clinical Features of LAM

The main clinical manifestations of LAM include pneumothorax, progressive dyspnea, and Chylous pleural effusions. More than 50% of LAM patients experience pneumothorax with 70% chances of recurrence (Young et al., 2006). Dyspnea is the

most common feature of LAM that develops in over 70% of these women and becomes universal as disease further progresses (Johnson et al., 2002). The other common symptoms include Chylous pleural effusions, cough, chest pain, hemoptysis, chylopysis, and wheezing (Johnson et al., 2002). Fatigue is one of the symptoms that has been overlooked even though it does present itself more often. In a cross-sectional study, Cohen et al. (2005) investigated two large patient registries to get a comprehensive update on the clinical features of LAM. They reported fatigue in 72% of their study population, elaborating on the fact that more women without prior episode of pneumothorax experienced fatigue and dyspnoea than those who had gone through such an episode in the past. Pneumothorax is a serious and frequent complication of LAM and it can be a leading factor causing death. Most common approaches to the treatment of pneumothorax have been pleural interventions such as pleurodesis, pleural abrasions, and partial pleurectomy (Young et al., 2006). Due to the possible serious complications and the negative consequences for the transplant candidacy that could be caused by these procedures, deciding on appropriate treatment option to prevent the recurrent episodes has been very challenging (Young et al., 2006). In an investigation looking at the opinions of pneumothorax treatment among 615 LAM foundation patient databases, Young et al. (2006) stated that the most patients favored a conservative approach when they were asked about what was their choice for pneumothorax management. Young et al. concluded that " in conjunction with appropriate pain management, a better understanding of patients' perspectives will facilitate cooperative decision making and may ultimately improve clinical outcomes in LAM related pneumothorax" (p.1267). The

significant impact of decline in lung function was reported by 41% of the study

population and most worries were noticed among those who had experienced only one

pneumothorax as well as those who frequently worried about developing an episode

(Young et al., 2006). With LAM physicians choosing more of an aggressive pleurodesis

in their surgical approach to management of pneumothorax and most patients being

favorable towards conservative treatment options, Young et al. elaborated on the fact that

most patients were reluctant to choose a definitive treatment yet they agreed to the fact

that pleurodesis helps prevent recurrence of pneumothorax. It is apparent that women

with LAM could greatly benefit once they are knowledgeable and informed about the

possible options they have and when they are faced with making the best decision to treat

and possibly prevent the recurrence.

In a cross-sectional study of 165 workers with COPD and asthma; Boot et al.

(2005) investigated the associations between knowledge about management of the

disease and knowing the diagnosis correctly, sick leave, health complaints, adaptation to

functional limitations, and perceived control. They concluded that more knowledge about

the disease, about management, and about diagnosis were associated with fewer patient

complaints about health and better perception of their disease. Boot et al. (2005) reported

that their results showed association between high knowledge about how to manage the

disease and adequate adaptation, which meant accepting the consequences of their

chronic illness and limitations to their daily functions. Perceived control was also

associated with knowledge; Boot et al. (2005) agreed to the fact that patients with more

of external factors such as fate or chance would be less likely to seek knowledge about

their disease and found that to be somewhat problematic. Patients with chronic

respiratory disease should have knowledge about managing and having control in case

symptoms such as asthma attack arise. The perception that some powerful other such as

the physician will control their health would discourage gaining the right knowledge and

control over exact diagnosis. Boot et al. (2005) stated that perceived control over health

through gained knowledge would most likely affect behavior change.

Extra-pulmonary features of LAM include abdominal Lymphadenopathy which

affects 50% of the LAM patients and it is seen in the upper retroperitoneum and pelvis,

presenting itself as fluid-filled cystic masses known as lymphangioeleiomyomas. These

masses could cause bloating and abdominal pain and approximately affect 16% of the

LAM population (Johnson et al., 2002). Angiomyolipomas (AMLs) are present in 50% of

the LAM patients and could also be present in the liver and pancreas as well. AMLs are

benign tumors that are found in 40% of the sporadic LAM and 80% of the TSC LAM

(Avila et al., 1999). The asymptomatic AMLs are small in diameter (1.3 cm) and usually

do not causes trouble, however the larger AMLs could be painful and disruptive by

causing hemorrhage and haematuria and call for emergency treatment (Ayo et al., 2007).

Conservative treatment or no treatment has been recommended with cautious follow ups

through once or twice ultrasound measurements for the larger asymptomatic AMLs; and

safe surgeries such as embolisation for when symptomatic patients are experiencing

bleeding and are in need of preserving their renal function (Hsu et al., 2002). It could be

helpful for the LAM population to be knowledgeable about the options available to them

with conserving their kidney function as it best suites their particular case and not to prematurely go along with the presented therapy.

Diagnosis

Diagnosing a rare disease such as LAM is extremely challenging and mistaken due to the similarities LAM presents among other respiratory diseases such as COPD and asthma. The European respiratory society guidelines for the diagnosis and management of LAM (ERS) were developed to produce evidence based, consensus guidelines for diagnoses, assessment, and treatment of LAM (Johnson et al., 2010). The diagnosis of LAM is made when characteristic lung HRCT and lung biopsy is compatible with the pathological criteria for LAM, characteristics lung HRCT and presence of either angiomyolipomas of the kidneys, thoracic or abdominal Chylous effusions, lymphangioeleiomyomas or lymph node involvement by LAM (Johnson et al., 2010). McCormack and Young (2010) stated that chest x-rays could be misleading and not reliable to provide convincing clues to diagnose LAM. They argued that high resolution CT scan should be proper initiative for all women who are younger presenting with documented pneumothorax and dyspnea and thereafter if the diagnosis of LAM is suspected. According to McCormack and Young (2010), about 80% of the time HRCT-scan can provide an accurate diagnosis of LAM. Even though the ideal accuracy would call for more than approximate 80%; other efforts such as full history, smoking status, family history of TSC, abdominal CT-scan or ultrasound to look for any existing renal AMLs should be performed to assure a more accurate diagnosis.

Treatment

Currently there are no definite treatments for LAM and even though LAM predominately affects women during their reproductive years and become worst during pregnancy; Harari, Torre, & Moss (2011) stated that no objective evidence has been significant when medical or surgical hormonal strategies were induced by using Gonadotropin-releasing hormone (GnRH) analogues or bilateral oophorectomy. Estrogen-containing medication as well as any soy products should be avoided, and patients should be informed that pregnancy could exasperate LAM (McCormack & Young, 2008). The pulmonary and abdominal complications are treated mostly through conservative therapies; with pneumothorax, aspiration or chest drain is strongly recommended; however if the air leak continues surgical pleurodesis is performed (Harari et al., 2011). The smaller AMLs are not treated aggressively and are basically followed up annually through ultrasound; although in order to treat larger and symptomatic AMLs embolisation or nephron-sparing surgery is performed (Johnson et al., 2002). Bronchodilators are considered for those patients who respond positively to the treatment. Oxygen should be recommended to maintain the oxyhemoglobin saturation of more than 90% at rest, exercise, and to maintain oxygen saturation level during sleep (McCormack & Young, 2008). Reynaud-Gaubert as quoted in Zhang and Travis (2010) stated that lung transplantation is the only effective treatment available for late stage or advanced LAM and the expected survival years after lung transplant are one year at 79.6%, 2 year survival at 74.4%, 5 year survival at 64.7, and 10 year survival at 52.4%.

Theoretical Framework

The theoretical framework for this study was based on the trans-theoretical stages of the change model which was developed by Prochaska and DiClemente in 1970s. The TTM was the right fit for this study because it focuses on decision-making of the target population and their intentions to change behavior. Prochaska and DiClemente stated that individuals do not just change behavior quickly; they will take time and will go through stages to adopt and maintain desired behavior change. The six stages of change include precontemplation which is stage of not desiring any change due to the fact that individuals are blinded to the pros of their decision to change or adopt a healthy behavior and more think about the cons of change. The second stage of change refers to the contemplation which individuals are opening up to the idea of change and will start to think about possibilities of pros versus cons of behavior change. The third stage of change is preparation to make the change within the next thirty days. The fourth stage refers to the action taken by the individuals and their intention to keep their effort in moving forward with the adopted behavior change. The fifth stage of change is about maintaining the adopted change and individuals every effort to prevent relapse to the pervious stage and the final stage of the process is all about termination and no intentions to go back to their unhealthy behavior. The desired objective for the LAM patients would be better quality of life by going through the stages of change and adopting the KAPs that are more conducive to possibly improve their quality of life.

LAM Foundation (2013) embraces the importance of exercise and education program tailored to meet the patient needs that could improve exercise tolerance and

feelings of shortness of breath. Although there are no guidelines or any studies to show the effects of exercise and quality of life for LAM patients; the literature in regards to the effects of exercise on COPD patients strongly supports the role of education and physical activity. One of these recommended activities is pulmonary rehabilitation which revolves around helping patients how to breathe more easily and improving quality of life. According to Cleveland clinic COPD exercise and activity guidelines; a well put together program to regular exercise will provide many benefits for these target population. Some of these benefits include; improving their circulation and help their body to better use of oxygen, build energy levels to be able to do more activities without becoming tired or short of breath, strengthen heart and cardiovascular system, strengthen bones, help reduce stress, anxiety, and depression, improve sleep, and achieve a better overall mental and physical health. Since LAM patients major complaints are shortness of breath encouraging such programs to improve breathing could also promise the same quality of life for LAM patients.

In another population based cross-sectional study utilizing the TTM; Salehi et al. (2010) investigated the physical activity and it's determinants among 454 participants aged 60 and over. Salehi et al. specifically looked in to the decisional balance and self-efficacy of TTM. Decisional balance refers to an individual's balancing out the pros and cons of behavior change and self-efficacy relates to the perceived confidence in individuals ability to initiate the behavior change and follow through maintenance stage (Salehi, 2010). The authors assessed the participant's knowledge in regards to physical activity and reasons why it is recommended to exercise; perceived benefits, and

perceived barriers regarding physical activity. Salehi et al. (2010) stated that there was a significant relationship between perceived benefits and stage of change. Most of their participants were aware of benefits to physical activity and their main reasoning for exercise was the benefit of social health. Sallis, as quoted by Salehi et al (2010), stated that physical activity and routine exercise has been influenced by many other factors such as social, cultural, environmental, and psychological therefore it is reasonable to promote initiatives that focus on the factors that tend to uplift suggested behaviors. Salehi et al. (2010) study results showed self-efficacy and perceived benefits were much more noticeable than perceived barriers in the later stages of TTM; and alluded to the fact that these two variables were most effective predictors of physical activity within their study group. They stated that participants who were more knowledgeable were more receptive to physical activity and concluded that in order to encourage healthy behaviors more focus should be directed towards increasing knowledge, increasing the target population perception on benefits than barriers, and enhance self-efficacy. It could be very beneficial to encourage interventions that could emphasize variables that tend to mediate positive behaviors; encourage self-efficacy, social support, and perceived benefits for LAM patients. Such interventions could have a positive impact on women with LAM to adopt KAPs that are conducive to possibly altering quality of their life. Yang and Chen (2005) stated that TTM encourages behavioral modifications to become part of the daily behaviors of individuals implying such modifications as developing process in to their lives. Prochaska and Norcross, (1994) commented on the fact that varieties of other therapeutic theories are affective in making up the TTM such as the behavior theory,

cognitive theory, existence theory, experiential theory, gestalt theory, human essence

theory, human relationship theory, psychological dynamics, and holistic theory method.

Prochaska and DiClemente (1982) stated that along with the six stages of change there

are factors and processes that facilitate the underlying behavioral change. According to

Prochaska and DiClemente (1982) theses ten processes included consciousness raising,

dramatic relief, environmental reevaluation, self-reevaluation, social liberation, counter

conditioning, helping relationships, reinforcement management, self-liberation and

stimulus control. Yang and Chen (2005) utilized the TTM stages of change to examine

the exercise stage and process of change in patients with obstructive pulmonary disease.

They used a cross-sectional correlation research design to investigate the distribution of

stages of exercise change and the process of change used for each stages of behavior

change among their study population of 98 COPD patients. Yang and Chen (2005) stated

that the process of change mostly utilized by the study participants were counter

conditioning, self-liberation, and self evaluation. They concluded that more behavioral

process was embraced by the participants in action and maintenance stage; and those in

precontemplation hardly used cognitive and behavioral process of change. Yang and

Chen (2005) elaborated that COPD patients tend to find it difficult to keep up with the

change behavior and in order to keep the early stage participants motivated; support

groups and telephone follow up should be routinely done in order to encourage the

patient's self-care responsibility. The authors stated that in the maintenance stage

participants were aware and knowledgeable about the importance of exercise and illness.

It is important to apply the counter-conditioning, self-liberation, and self-reevaluation

strategies in order to discourage patients from regressing to the earlier stage. Encouraging and reinforcing the fact that such behavior change is significant in improving related health problems, and as such improved quality of life; is critical component to reach a desired outcome. Yang and Chen (2005) study results provided importance for the stage-matched interventions geared at exercise that could possibly help COPD patients maintain adopted behavior. According to the authors; initiating such assistance to these target population would reduce the worsening of pulmonary function, allows for keeping up with the daily activities, and most of all quality of life. Yang and Chen(2005) concluded " efficient self change depends on doing the right things (processes) at the right time (stages), and also encouraging the family members to participate in exercise plan to maintain patients exercise behavior" (p. 102). There are no studies done on the effects of behavior change to improve LAM related health problems. Utilizing the TTM through incorporating cognitive and behavioral process of change as well as applying strategies such as counter-conditioning, self-liberation, and self-reevaluation, could possibly benefit LAM patients. Adopting the stages of change and KAPs would aim these target population in improving their knowledge of LAM, therefore, embracing behavior and lifestyle practices that could ultimately improve their quality of life. LAM is mostly misdiagnosed due to the lack of disease awareness among health care professionals and similarity it presents to the other lung diseases such as asthma or emphysema (S. Johnson et al., 2002). It is apparent that despite efforts by the LAM foundation and other LAM organizations; patients are being misdiagnosed and affected by delayed diagnosis. It is very challenging to identify such a rare disease when the similarities are much too close

to other obstructive diseases such as COPD or asthma which happen to be less morbid

than LAM (McCormack & Young, 2010). Gaining knowledge and understanding of this

disease can not only be critical for health care communities but also could be essential for

women who have been identified with this life threatening disease. Lua and Neni (2011)

studied the importance of awareness, knowledge, attitude and its importance in quality of

life of people with epilepsy; and concluded that incorrect knowledge and awareness

caused more suffering that the disease itself. In their cross-sectional study in regards to

the awareness, knowledge, and attitude (AKA) of patients with epilepsy and their quality

of life; Lua, and Neni (2011), reported that patient's awareness about their disease greatly

correlated with the seizure worry and incorrect knowledge was a variable negatively

affecting seizure worries. Lua and Neni (2011) stated that attitude was greatly associated

with good mental state and cognitive behaviors, social activities and increased energy.

They eluded that knowledge and awareness along with positive attitude had a noticeable

relation to the patient's quality of life; and as result of their findings Lua and Neni

(2011), concluded that it is essential to initiate educational initiatives to continuously

improve on AKA levels to impact quality of life.

In another study, Reema, Adepu & Sabin (2010) investigated the impact of the

KAPs on the therapeutic outcome of 27 patients with chronic obstructive disease

(COPD). They initiated an intervention examining the baseline KAPs of these patients in

regards to their disease which showed patients poor knowledge of COPD as well as low

attitude and life style practice. Reema et al. (2010) stated that more than 50% of the

patients thought COPD is curable and few thought of smoking as not a factor in

deteriorating their disease. 20% of the patients did not put importance in taking their

medication properly and thought medicine is only needed as they experience acute

exasperations. Reema et al. (2010) elaborated that knowledge is a variable that may be

useful in how patients feel about their disease and how to best manage their condition.

They identified in their study that increased knowledge and motivation reflected on the

way patients perceived their COPD and pointed out that the two factors enhancing

treatment outcome; (a) would be patient counseling in order for better understanding of

treatment options, (b) to motivate patients through therapy adherence. Reema et al.

(2010) concluded in their findings that at the end of study; 65% of the participants

responded that smoking is critical risk factor in worsening of their COPD and 90%

practiced scheduled medication use. Comparing to the baseline knowledge of ease of

inhaler use by the 11% of the study participants; at the end of the study 80% of the

participants were knowledgeable about the ease of inhaler use.

It is crucial to understand the chronic disease characteristics and factors that could

dramatically affect the patient's health overall. Holman and Lorig (2004) stated the five

categories of outcome that are aimed for by the health care providers include:

"physiology, symptoms, physical and emotional function, personal health perceptions,

and quality of life" (p.119). They eluded that functional ability did not appear significant

until late 1960s and 1970s when the chronic diseases starting to rise continually. Holman

and Lorig (2004) elaborated that measuring functional capacity became relevant in newly

designed instruments which not surprisingly proved other variable to be just as

significant. Chronic diseases take a tremendous toll on the patient's lifestyle, and their

attitude and knowledge of their disease has great significance in the patient's functionality. According to Holman and Lorig (2004) the patient's quality of life and survival were much better if the patient's had positive attitude and strived to overcome the concerns caused by their chronic condition. LAM is a debilitating disease that can potentially suffocate women (LAM Foundation, 2013). Enhancing Knowledge and promoting positive attitude can be conducive to LAM patients understanding and managing their chronic disease as it has been shown to be effective in above mentioned studies investigating the same objectives.

In a cross-sectional assessment of KAPs among Hepatitis-B patients in Quetta, Pakistan, Haq et al. (2013) concluded that the patients attitude in regards to their disease had a significant impact on their performed behavior towards HB. The authors further elaborated that "The attitude was shaped because of the knowledge that patients possesses regarding HB infection hence it is concluded that correct knowledge brings about a positive attitude and this positive attitude brings about a positive change in the practices of the patients" (p.7). Kanaka Durga Devi and Hema Suma Sree (2012) investigated the importance of KAPs and its relation to disease and its management in patients with asthma. They assessed the KAPs of the study participant at the baseline and again at the final follow up. Patient education was developed and provided to the patients in order to increase their knowledge about the disease, medications, and lifestyle modifications. Kanaka Durga Devi and Hema Suma Sree (2012) study results suggested that increased KAPs, through education improves patient's ability to understand the symptoms of their disease as well as gaining ability to make informed decisions about

their therapies. They also elaborated on the fact that one of the key factors about patient education and knowledge is to recognize the risk factors triggering the symptoms of their disease; these factors can be smoking, dust mites, outdoor air pollution, insect's allergen, and mold (Kanaka Durga Devi & Hema Suma Sree, 2012). Due to the rarity of LAM proper diagnosis and management of this disease is very challenging, therefore it makes it more critical for patients to become knowledgeable and aware of symptoms and possible risk factors. In order to gain a better perception on how to overcome the uncertainty they may have with identifying related symptoms, LAM patients have to improve their knowledge and change behavior in order to possibly impact quality of life.

LAM is a chronic disease that is progressive and has no cure. Women who are newly diagnosed with this debilitating disease are scared and in state of disbelief at the time of diagnosis. For so many receiving the diagnosis of LAM causes anger, denial, shock, grief, helplessness, confusion, despair, sadness, and fear (The LAM Foundation, 2013). For many this news could also validate that what they are experiencing has reasons and explanation has a name (The LAM Foundation, 2013). Patients have to confront their illness and becoming knowledgeable about the facts and myths of LAM could allow women to change behavior and lifestyle modifications in order to possibly improve their quality of life.

In another study, Leino-Kilpi et al. (2005) analyzed the relationship between patient education and quality of life among 237surgical hospital patients in Finland. Based on their findings it appeared to be association between acquired knowledge and gained quality of life. Leino-Kilpi et al. (2005) showed the higher the quality of life index

was indicative of the knowledge the patient had. They also compared the two groups; 15% of the patient with best quality of life and 15% of the patients with worst quality of life and determined that the group with best quality of life had higher knowledge and higher education than the group with worst quality of life. Leino-Kilpi et al. (2005) concluded "subjective health-related quality of life and received knowledge correlate and vice versa" (p.313). In a cross-sectional study; Mancuso, Sayles & Allegrante (2010) studied the knowledge, attitude, and self efficacy in asthma self management and quality of life in 180 patients with the mean age of 43. Mancuso et al. (2010) objective was to investigate what clinical and patient characteristics were associated with the cognitive variables within the three domains of knowledge, attitude, and self-efficacy asthma questionnaire. They also assessed contributions of these variables to different domains of clinical status measured by the asthma quality of life questionnaire (Mancuso et al., 2010). Over 50% of their study population had low knowledge in regards to their disease, fair attitude and moderate self-efficacy. Correlation between knowledge and attitude as well as knowledge and self-efficacy were lower than correlation between attitude and self- efficacy among the study population which in turn negatively affected the quality of life of asthma patients (Mancuso et al., 2010). Other variables such as social support and less depressive episodes were associated with more positive attitude and motivation to maintain their adopted behavior towards asthma acceptance (Mancuso et al., 2010). The authors concluded that knowledge of asthma is critical for the individuals to be self managers; to understand their disease and be able to monitor their lung function,

recognize exacerbations early, dose rescue medications, and being able to recognize when emergency care at the hospital is needed (p.5).

Although there are no studies looking at the importance of KAPs of LAM patients and how it affects their quality of life; the above literature review points out many findings significant to improving quality of life in patients with chronic disease especially lung diseases such as COPD and asthma. It is therefore imperative to initiate studies investigating the importance of interventions encouraging KAPs and adopting behavior to impact quality of life for women with LAM.

Summary and Transition

This chapter consisted of literature review related to the role of KAPs in chronic diseases and how it relates to their quality of life. The objective of this study was to look at KAPs in LAM patients and how they related to their quality of life; however, due to the lack of any literature in this regards I have discussed studies with similar objectives. This chapter also covered clinical data on LAM and its etiology: types of LAM, clinical presentations, pulmonary physiology, diagnosis, and available treatment. This review also presented information in regards to the importance of TTM and stages of change; also studies presenting the effects of self-efficacy and self-management in adopting the behavior change in COPD and asthma patients. The association between knowledge and quality of life was also discussed. This chapter concluded by mentioning the need for studies to investigate the importance of initiatives promoting KAPs conducive to the possible altering quality of life in patients with LAM. The next chapter will cover the methodology section.

Chapter 3: Research Method

Introduction

This chapter will include the detailed description of the research design and will cover the target population, instrumentation, data collection and analysis; this section will also give an overview of the rationale for selecting such a design for this particular study. The purpose of this study was to investigate the knowledge, attitude, and the lifestyle practices (KAPs) of the women with LAM and how these variables affected their quality of life.

Research Design and Rationale

The focus of this study design was to understand the effects of KAPs on quality of life of women with sporadic LAM. In order to examine the relationship between these variables and quality of life as the dependent variable; a quantitative study utilizing a survey design was selected as the appropriate approach for the present study. Due to the gap in the literature there have not been any KAPs questionnaires designed specifically for the LAM population. In order to measure the independent variables of knowledge, attitudes, and lifestyle practices, KAPs questionnaires validated for a similar disease such as chronic obstructive pulmonary disease (COPD) were used to study the sample of the sporadic LAM population (see Appendices D, E, F, G). A quality of life survey questionnaire was employed to measure the dependent variable; in this case the quality of life among study participants (see Appendix H). According to Creswell, (2009), using a quantitative approach will facilitate a numeric explanation of opinions of a study population which then makes it possible for the researcher to make a certain claim on that

particular sample population. The quantitative study design did appropriately allow for the key questions designed for this study to examine the KAPs of the LAM patients within the study sample population. This study approach measured the KAPs variables and determined the relationship between the KAPs of this LAM sample population and the quality of their lives.

According to the literature , although there has not been any research done in regards to the KAPs variables and their relationship to the quality of life within the LAM population; there have been several quantitative studies examining the KAPs of patients with other chronic diseases. In regards to the affects of KAPs in a chronic disease condition, it is important to look at other existing literature indicating the relationship between KAPs and quality of life. In a cross-sectional study, Lua and Neni (2011) looked at the importance of awareness, knowledge, attitude and its importance in quality of life of people with epilepsy. They concluded that incorrect knowledge and awareness caused more suffering than the disease itself. Lua and Neni (2011), reported that patient's awareness about their disease greatly correlated with the seizure worry and incorrect knowledge negatively affected seizure worries. Lua and Neni (2011) stated that attitude was greatly associated with good mental state and cognitive behaviors, social activities and increased energy. They concluded that knowledge and awareness along with positive attitude had a noticeable relation to a patient's quality of life; therefore, it is essential to initiate educational initiatives to continuously improve on knowledge and attitude to impact quality of life. In another study, Reema, Adepu, and Sabin (2010) investigated the impact of the KAPs on the therapeutic outcome of 27 patients with chronic obstructive

pulmonary disease (COPD). They examined the baseline KAPs of these COPD patients, which showed patients' poor knowledge of COPD as well as low attitude and lifestyle practice. Reema et al. (2010) stated that more than 50% of the patients thought COPD is curable and few thought of smoking as a factor in deteriorating their disease. Twenty percent of the patients did not put importance on taking their medication properly and thought medicine was only needed as they experience acute exasperations. They discovered in their study that increased knowledge and motivation reflected on the way patients perceived their COPD and pointed out that the two factors enhancing treatment outcome would be patient counseling to have a better understanding of treatment options; and to motivate patients through therapy adherence.

Setting and Sample

The patient population in this study was a sample of 143 women with sporadic LAM. Based on the power analysis that was done utilizing G power 3.1 to determine the effect size of .10 with a power of .80 including 10% for replacement, a size of 134 participants was determined to be adequate in order to achieve an alpha level of .05. The participants who took part in this study were registered with the LAM Foundation that has the largest number of LAM patients registered to date (LAM Foundation, 2013). This allowed for ease of accessibility with sampling strategy. Permission was obtained from the director of the patient services at LAM Foundation for the purpose of facilitating access to the participants by sharing the link to the survey questionnaires. (see Appendix A)

The Inclusion Criteria

These inclusion criteria for study participants were (a) diagnosed with sporadic LAM; (b) fluent in English language; and (c) age of more than 18 years.

The Exclusion Criteria

These exclusion criteria for study participants were (a) not diagnosed with sporadic LAM; (b) not fluent in English language; and (c) not more than 18 years of age.

Procedures

For the purpose of data collection an account was created with Survey Monkey for distributing the questionnaires to the participants as well as receiving the responses back as they were completed. The study flyer with invitation and direction to the link was posted for the participants. An informed consent page was created and sent through Survey Monkey to the participants to read before they proceeded with the questionnaires (see Appendix I). The Survey Monkey created a unique web address for the survey questionnaires along with the contact information. The e-mail address as well as phone numbers was included in the contact information in case of any concerns or questions participants had throughout their participation in the study. Survey Monkey assigned a unique number for each participant for when she completed the questionnaires and the IP addresses were recorded in order to prevent duplication. Once the informed consent was signed by the participants, they had an opportunity to fill out a demographic questionnaire that inquired about age, education, ethnic background, and income followed by the KAPs and the quality of life questionnaires. Once all the surveys needed were collected, a request for a data file of each respondent's answers was sent in to

Survey Monkey. Once all the answers were received, the data were exported to SPSS for analysis.

It was brought to participants' attention that the findings of the study could be shared with any participants who liked to receive the study's results

Instrumentation

A demographic questionnaire assessed the study participants' age, ethnic background, education, and income.(see Appendix G) A KAPs questionnaire consisting of the Bristol COPD Knowledge Questionnaire (BCKQ); (White, Walker, Roberts, Kalisky, & White, 2006; see Appendix D); the Beliefs and Behaviour Questionnaire (BBQ); (Johnson, Mackinnon, Kong, & Stewart, 2006; see Appendix F); the Determinants of Lifestyle Behavior Questionnaire (DLBQ); (Lakerveld et al., 2011; see Appendix E); and the St George's Respiratory Questionnaire (SGRQ); (Jones, Quirk, & Baveystock, 1991; see Appendix H) were accessed through an e-mail provided link to Survey Monkey to the participants, which then measured the KAPs of these target population. The BCKQ was developed by Dr Roger White and validated in order to assess the knowledge of individual patients with COPD. White, Walker, Roberts, Kalisky, and White et al. (2006) stated that BCKQ embodies a good reliability and internal consistency that examines the knowledge related to the COPD patients. The BCKQ is a multiple-choice questionnaire that consists of 13 topics. Each question contains five statements that could be answered *true, false* or *don't know* (White et al., 2006). Positive scoring (+1) was given for a *true* answer and no point (0) was given to

the *false* or *don't know* response. Permission to use this questionnaire was granted from Dr White (see Appendix B)

The BBQ was design and validated to assess the health beliefs and behavior adherence in patients with chronic diseases. This questionnaire contains 30 items and the measurement is on a five-Point Likert-type scale (G. Johnson et al., 2006).

The DLBQ was validated to assess the determinants of lifestyle behavioral change with three components of diet, physical activity, and smoking (Lakerveld et al., 2011). This questionnaire consists of 50- items on attitude, subjective norms, perceived behavioral control, and intentions. The attitudes were measured on a 7 point semantic scale ranging from 1 being unpleasant to 7 being pleasant and the other determents of change were measured based on the 5-point likert-type scale ranging from 1 totally disagree to 5 totally agree. (Lakerveld et al., 2011)

The SGRQ that was utilized in this study to measure the quality of life variable was developed by professor Paul Jones in 1991 It is a self-administered health related quality of life questionnaire for patients with respiratory health diseases. The SGRQ is a 50- item instrument that incorporates the three categories of symptoms, activity, and impact. The subscale of symptoms includes 8 items which covers respiratory symptoms and their severity, the activity subscale which includes 16 items focuses on breathlessness and the activities that are limited or the cause of breathlessness, and impacts which includes 26 items concentrating on social functioning and psychological effects as results of respiratory problems (Jones et al., 1991) The scoring ranges from 0%-100% ; zero indicating no impairment of the quality of life, and it is calculated for each subscale as

well as the total score. This scoring is based on the weights as an estimate of the distress for each item of the questionnaire. Permission to use the SGRQ was received from the developer of the instrument. (See Appendix G)

Due to the rarity of LAM there has been no diseases specific tools developed to measure the quality of life among the LAM population. The St George's respiratory questionnaire (SGRQ) is the only respiratory specific health related quality of life tool that has been examined in LAM related studies. Swigris et al. (2013), examined the SGRQ longitudinal construct validity in LAM Analysis was performed to determine the relations between the SGRQ scores and values for four external measures (Swigris et al., 2013). The data used in the study were pulled from the Multicenter International Lymphangioleiomyomatosis Efficacy and Safety of Sirolimus trial. The authors measured the FV1, diffusing capacity for the carbon monoxide of the lungs, the 6-min walk test and the distanced walked, and the serum vascular endothelial growth factor-D (VGEF-D) Swigris et al. (2013) concluded that SGRQ scores were correlated with the measurement values at the baseline as well as 6month and 12 months evaluations. Their longitudinal analysis results indicated that the SGRQ change score also tracked changes in values for FV1, diffusing capacity,6-min walk test, and the VEGF-D Swigris et al. 2013, concluded that at 12 months the participants with the improvements in their scores showed higher improvements in their SGRQ scores their results of cumulative distribution also showed further support for the longitudinal validity of the SGRQ in LAM. Xu et al. (2010) authored the other study that looked at the SGRQ in LAM. They examined the correlation between the health related quality of life measured by SGRQ and the

physiological measures in LAM. Xu et al. (2010), analyzed the SGRQ scores and other measures such as 6-min walk, oxygen levels in blood, and pulmonary function test (PFT) in patients with LAM .They concluded that the mean values of the three components of the SGRQ symptoms, activity, and impact as well as the total scores were correlated with the Borg scale of breathlessness which included the 6-min walking test, oxygen level in the blood, the PFT and the diffusion capacity of the lung. Xu et al. (2010) elaborated that in their preliminary observation sirolimus improved the SGRQ total and the three component scores in symptoms, activity, and impact as well as the Borg scale of breathlessness. The conclusion of this study stated that the SGRQ can be recommended as baseline and follow up evaluation with patients with LAM (Xu et al., 2010)

Analysis

This study utilized SPSS in order to do the analysis. Descriptive statistic was employed to describe the sample population and the collected data. Pearson's correlation was utilized for each one of the independent variables; KAPs, in order to examine each variable's relation to the quality of life. Finally, linear regression was used to predict quality of life from KAPs, controlling for demographic variables. Data was examined for outliers prior to analysis. The research questions as well as the hypothesis are restated for review.

Research Question 1: To what extent does level of knowledge about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 1: There is an influence of the knowledge about LAM in women living with LAM on their quality of life.

Null Hypothesis 1: There is no influence of the knowledge about LAM in women living with LAM on their quality of life.

Research Question 2: To what extent does the attitude about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 2: There is an influence of the attitude about LAM in women living with LAM on their quality of life.

Null Hypothesis 2: There is no influence of the attitude about LAM in women living with LAM on their quality of life.

Research Question 3: To what extent do the life style practices of women living with LAM influence their quality of life?

Alternative Hypothesis 3: There is an influence of life style practices of women living with LAM on their quality of life.

Null Hypothesis 3: There is no influence of life style practices of women living with LAM on their quality of life.

Table 1

Variables and Statistical Tests per Research Question

Research Question	Variables	Statistical Test
Question 1	DV: SGRQ score IV: B CKQ score	Pearson's Correlation
Question 2	DV: SGRQ score IV: BBQ score	Pearson's Correlation
Question 3	DV: SGRQ score IV: DLBQ score	Pearson's Correlation

Education and income were confounding variables and an attempt was made to remove their influence before looking at the independent variables affect on the quality of life. The threats to the external validity are the fact that study participants were randomly

selected from only one patient's registry and the results will only serve the population registered with that particular registry and not generalized among the rest of the LAM population elsewhere. In regards to the threats to internal validity; since this study was taking place within a short period of time the possible threat from history was slim and since the study participants were only taking at one time and not repeatedly over time; the maturation threat to internal validity was also unlikely.

Ethical Procedures

Due to the nature of the study and ethical consideration the participants were provided with the information to fully explain the nature of the study. This information covered the confidentiality issues, risk and benefits, the detailed explanation on the study, the step by step procedures for the participants to follow, explanation on the importance of confidentiality and the ethical concerns, as well as emphasis on the absolute voluntarily participation in the study. E-mail address as well as phone numbers was provided in case of any concerns or question participants had throughout their participation in the study.

Every consideration was taken to reassure the participant's full understanding of confidentiality, and ethical concern by explaining in detail that their information would be kept confidential, and the only one having access to their data would be the researcher. The participants who sign the consent were also notified of their will to pull away from the study at any time and the facts in regards to no risks or benefits for the participants were also emphasized. On June 12, 2014 approval from the Walden University Internal

Review Board (IRB) was received in order to conduct the present study with the approval

number of 06-13-14-014-0145430.

Summary and Transition

In this chapter the proposed study discussed the details of the research design and

the methodology that was selected to collect new data. The complete description of every

step including the sample selection, instrumentation, procedure, and data analysis were

given as to how the relationship between KAPs and the quality of life of women with

LAM were examined. Based on a power analysis utilizing G power 3.1; 134 participants

were needed to make an effects size of .10 with a power of .80. The sampling process for

the present study was selection of 143 women with sporadic LAM. Procedures for data

collection was done through an online survey; facilitated by Survey Monkey that

provided a link for the participants to get the questionnaires. An informed consent had to

be signed before participants could proceed with their responds to the questionnaires.

Employing the BCKQ, the DLBQ, the BBQ, and the SGRQ instrumentation had been

validated for use as tools measuring the knowledge; attitude, practices, as well as quality

of life .Pearson correlation and linear regression were utilized as the valid statistical tests

selected to examine the variables and research questions. The ethical concerns

information covered the step by step procedures for the participants to follow;

explanation on the importance of confidentiality and the absolute voluntarily participation

in the study were emphasized in great deal.

The following Chapter 4 will describe the data analysis methods, will summarize

the results of the statistical analysis, and will present the major findings. This chapter will

discuss the sample population, it will address the research questions and the hypothesis, as well as tables in order to illustrate results It was expected that the findings of this study would provide new data, filling the gap that exist in the current literature in regards to identifying the affects of KAPs and the quality of life in LAM patients.

Chapter 4: Results

Introduction

The purpose of this study was to look at the KAPs of the LAM patients and how they affected their quality of life. A sample of 143 LAM patients participated in the KAPs and quality of life survey questionnaires. Three research questions and related hypotheses were examined in order to find out the relationship among the three independent variable; knowledge, attitude, and lifestyle practices and the dependent variable quality of life. Data analysis was done to determine the association. This chapter will explain the analysis of collected data including the descriptive and demographic characteristics of the study population, the results and the statistical analysis, and the tables and figures in order to illustrate results.

Data Collection

Data were collected between July 1, 2014 and August 25, 2014 on Survey Monkey. Utilizing The LAM Foundation website; the study flyer with invitation and direction to the link was posted for the participants. Due to the request of the LAM Foundation's executive director (S. Sherman, personal communication, July 2014), I was directed to post the study information as well as the invitation reminders on the LAM Foundation's Facebook page. A reminder was posted on a weekly basis to reassure further recruitment.

As of August 25, 2014, 171 opened the survey and 164 consented. Of those 164, 21 answered no questions, so were considered to have refused the survey. The remaining 143 respondents were considered the sample for this study. Recruitment rate was 13.8%

and the response rate for those who viewed the survey was 83.6%. Note that a sample size of 134 was determined to be adequate to produce power of 0.80 with a small effect size. The target sample size was 147 (10% above the 134 needed). The sample size obtained was above 95% of the target sample size.

The data collection proceeded as planned. Based on feedback from the first set of respondents, a change in how the online survey worked was made. Originally, all questions had to be answered. One section of the DLBQ on smoking was only appropriate for those who smoked and could not accurately be answered by non-smokers. A change was made to make all questions optional, except the consent. The sample population for this study was made up of women with sporadic LAM who were members of The LAM Foundation. Table 2 displays the demographic breakdown of the sample.

Table 2

Frequency Distribution of Socio-demographic Characteristics of Participants (N=143)

	Frequency	%
Age		
25-34	9	8.3
35-44	26	24.1
45-54	47	43.3
55-64	22	20.4
65-74	4	3.7
Hispanic, Latino, or Spanish Origin		
No	103	95.4
Mexican, Mexican American, Chicano	1	.9
	2	1.9
Puerto Rican		
	2	1.9
Other Hispanic		
Race		
Caucasian/White	99	92.5
Black/African American	3	2.8
Chinese	1	.9
Filipano	1	.9
American Indian/Alaska Native	1	.9
Education		
Less than high school	1	.9
High school graduate	20	18.7
Associate degree	10	9.3
Bachelor's degree	44	41.1
Master's degree	29	27.1
PhD or professional degree	3	2.8
Household Income		
10,000-19,999	7	6.9
20,000-29,999	5	5.0
30,000-39,999	5	5.0
40,000-49,999	12	11.9
50,000-59,999	8	7.9
60,000-69,999	4	4.0
70,000-79,999	11	10.9
80,000-89,999	6	5.9
90,000-99,999	10	9.9
100,000 or more	33	32.7

Note. The response rate for online surveys overall is 13.35% (Hamilton, 2009).

Results

Age

Age was recorded into three groups of more homogenous sizes than the original

5: 25-44 years, 45-54 years, and 55 years and older. Table 3 presents the descriptive

results of SGRQ scores by age group. One-way ANOVAs indicated differences by age

only on the Activity subscale $F(2, 101) = 3.18$, $p < .05$; post hoc testing using Bonferroni

indicated that the difference was between those 25-to-44 years old those 45-to-54 years

old, with the younger age group having lower Activity scores indicating fewer impacts on

their activities.

Table 3

Means and Standard deviations for St. George's Respiratory Questionnaire (SGRQ)

(Total and Subscales) by Age Group

Age	SGRQ Total M (SD) n	Symptoms M (SD) n	Activity M (SD) n	Impacts M (SD) n
25-44 years	40.7 (22.3) 24	46.7 (22.7) 24	47.8* (28.9) 34	29.4 (20.9) 34
44-54 years	50.3 (19.9) 31	53.9 (24.0) 31	64.1* (24.7) 46	36.9 (19.7) 45
55 years and older	46.3 (21.2) 14	49.4 (26.5) 15	60.9 (27.4) 24	31.8 (20.5) 21

*$p < .05$.

Income

Income was recorded into 2 approximately equal sized groups: $10,000-$69,999,

and $70,000 or more. Table 4 presents descriptive results of SGRQ scores by recoded

income groups:

Table 4

Means and standard deviations for St. George's Respiratory Questionnaire (SGRQ)

(total and subscales) by Income Group

Income Level	SGRQ Total M (SD) n	Symptoms M (SD) n	Activity M (SD) n	Impacts M (SD) n
Below $70,000	55.0*** (20.2) 30	58.6* (21.4) 31	70.4** (21.2) 40	43.1*** (20.1) 40
$70,000 or more	38.9*** (19.6) 30	43.7* (24.5) 38	53.1** (27.6) 58	27.1*** (17.8) 55

Note. Independent samples t-tests found significant differences in mean SGRQ total ($t(67) = 3.32$, $p = .001$) and all subscale scores (Symptoms $t(67) = 2.67$, $p = .01$; Activity $t(94) = 3.52$, $p = .001$; Impacts $t(67) = 4.11$, $p < .001$) between the two income groups. *$p < .05$. **$p < .01$. ***$p < .001$.

Education

Because only one person had less than degree high school diploma and two had

PhDs, education was reduced to the following four groups: high school graduate or less,

associate's degree, bachelor's degree, master's degree or higher. Table 5 presents the

mean SGRQ scores by education level. One-way ANOVA revealed no significant

differences by education groups in SGRQ total score or any of the subscale scores.

Table 5

Means and standard deviations for St George's Respiratory Questionnaire (SGRQ)

(total and subscales) by Education Group

Education Level	SGRQ Total M (SD) n	Symptoms M (SD) n	Activity M (SD) n	Impacts M (SD) n
HS or less	41.4 (21.9) 13	43.9 (20.8) 13	48.5 (28.4) 21	33.1 (20.8) 20
Associate degree	56.6 (9.0) 8	64.3 (11.1) 8	71.3 (28.7) 10	39.2 (17.0) 10
Bachelor's degree	44.2 (22.3) 29	49.8 (26.9) 30	60.7 (25.4) 42	33.3 (20.1) 40
Master's degree or higher	47.7 (22.8) 18	49.8 (24.9) 18	56.8 (28.6) 30	30.8 (22.0) 29

Quality of Life

The quality of life was measured using the SGRQ, which produces an overall

quality of life score and three subscale scores—symptoms, activity, and impacts. The

higher the total score, the more the disease adversely affects quality of life. Similarly, the

higher the score for symptoms subscale indicates more severe symptoms; the higher the

score for activity indicates a greater effect of the disease on daily activities; and the

higher the score on impacts subscale indicates more severe impacts on quality of life.

The means and standard deviations for the total SGRQ and all subscales are presented in

Table 6 for those participants who provided enough answers to score the SGRQ; it is

obvious from Table 5 that their scores are much higher than people without respiratory

illnesses. One-sample t-tests were used to compare the scores on SGRQ subscales and

total to those for a group of people without respiratory disease (Jones et al., 1991). On

the Symptoms subscale, the LAM sample had a significantly higher mean than the

normal sample (M = 12), $t(69) = 13.42$, $p < .001$. The LAM sample was significantly

higher on the other subscales and the total, as well: for Activity (normal sample mean of

9), $t(103) = 18.22$, $p < .001$; for Impacts (normal sample mean of 2), $t(99) = 15.38$, $p <$

.001; and for total SGRQ score (normal sample mean of 6), $t(68) = 15.75$, $p < .001$

Table 6

Mean scores on St. George's Respiratory Questionnaire

	N	Mean	SD
Symptoms	70	50.5	24.0
Activity	104	58.1	27.5
Impacts	100	33.3	20.4
Total	69	46.1	21.2

Beliefs and Behaviors

The Beliefs and Behaviors Questionnaire (BBQ) was used to assess three

domains: beliefs about illness and treatment, experiences with the disease, and behaviors

related to adherence to treatment. Each domain has a positive and negative subscale.

The means and standard deviations for the BBQ domain subscales and individual items

are displayed in Table 7. Higher scores indicate higher levels of agreement with

individual statements (rated on a 1 to 5 scale). On the whole, this sample is about equally

confident (M = 3.77) and concerned (M = 3.45) about their illness management. They

are relatively satisfied (M = 3.98) and not disappointed (M = 2.85) with their doctors and

treatment, and are more adherent (M = 3.90) than nonadherent (M = 2.59) to their

medicine and treatment regimens.

Table 7

Means and Standard Deviations by Domain Subscales and Items for Belief and Behaviors

Questionnaire

	N	Mean	SD
Beliefs			
Confidence Subscale	143	3.77	0.68
I have sufficient understanding about my illness	142	3.99	0.99
I know what to expect from my illness management.	143	3.72	0.97
My current management will keep my illness at bay.	142	3.06	0.91
I am receiving the best possible management.	142	3.76	1.02
The management of my illness is a mystery for me.[a]	143	2.13	1.04
My medications are working.	137	3.47	0.85
I have a say in the way my illness is managed.	142	4.18	0.72
I have sufficient understanding about the options for managing my illness.	143	4.03	0.93
My doctors are very knowledgeable.	143	3.86	1.14
Concerns Subscale	143	3.45	0.39
It is helpful to know the experiences of others with similar illness as mine.	142	4.46	0.67
Natural remedies are safer than medicines.	143	2.68	0.87
My doctors have limited management options to offer me.	141	3.34	1.16
Using any medication involves some risk.	143	4.26	0.72
I am on too many medications.	138	2.48	0.96
Experiences			
Satisfaction Subscale	143	3.98	0.80
My doctors are compassionate.	141	4.23	0.79
I am satisfied with the information my doctors share with me.	142	3.73	1.00
My doctors spend adequate time with me.	143	3.97	1.07
Disappointment Subscale	143	2.85	0.72
I am concerned about the side effects from my medications.	137	3.40	0.98
Financial difficulties limit my access to the best healthcare.	143	2.38	1.17
The management of my illness disrupts my life.	142	3.05	1.13
Behavior			
Adherence Subscale	140	3.90	0.62
I have strict routines for using my regular medications.	136	3.90	0.90
I keep my medications close to where I need to use them.	135	4.12	0.72
I ensure I have enough medications so that I do not run out.	136	4.12	0.75
I push myself to follow the instructions of my doctors.	140	3.56	0.90
Nonadherence Subscale	142	2.59	0.69
I vary my recommended management based on how I am feeling.	136	2.69	1.14
I put up with my medical problems before taking any action.	141	2.82	1.12
I get confused about my medications.	136	1.85	0.78
I make changes in the recommended management to suit my lifestyle.	138	2.96	1.06

Knowledge

Knowledge of respiratory illness was assessed using the Bristol COPD

Knowledge Questionnaire (BCKQ) which assesses knowledge in 13 different areas as

well as providing an overall knowledge score. Table 8 shows the mean percentage of

correct responses to the total instrument as well as to each of the 13 subsections

Table 8

Knowledge Questionnaire (BCKQ) by Topic

Topic	Average Total
1 Epidemiology	58.4
Topic	Average Total
3 Symptoms	49.3
4 Breathlessness	51.7
5 Phlegm	53.6
6 Infections	55.3
7 Exercise	74.6
8 Smoking	69.3
9 Vaccination	77.8
10 Inhaled bronchodilators	55.7
11 Antibiotics	62.5
12 Oral steroids	48.8
13 Inhaled steroids	19.5
Total knowledge score	54.5

Note: Each topic had five questions. Actual questions and results for individual items can be found in Appendix K. "F" correct response was False; "T" correct response was true. Don't know is considered an incorrect response in calculating percentage correct.

The higher the percentages correct the more knowledgeable. The sample's

knowledge was highest for vaccination, exercise and smoking, and very poor on inhaled

steroids. Overall, the sample was correct 54.5% of the time, only slightly better than

chance responding which would be expected to produce a 50% correct response rate

Attitudes and Behavior

Attitudes about physical activity, healthy eating and smoking, and intentions to

change behavior in those three areas were assessed by the DLBQ. As can be seen in

Table 9, the majority of respondents had positive attitudes towards being more physically

active, eating healthy foods and, for those who answered the smoking section, stopping

smoking

Table 9

Percentages of Respondents by Attitudes Towards Healthy Behavior (DLBQ)

In my opinion:	N	Being More Physically Active	N	Eating Healthy Foods	N	Stopping Smoking
Pleasant	135	74.1	128	90.6	68	70.6
Unpleasant		25.9		9.4		29.4
Satisfactory	136	62.5	129	88.4	68	79.4
Frustrating		37.5		11.6		20.6
Good	134	98.5	128	97.7	68	98.5
Bad		1.5		2.3		1.5
Important	136	100.0	129	99.2	68	100.0
Unimportant		0.0		0.8		0.0
Desirable	135	94.8	128	96.1	67	100.0
Undesirable		5.2		3.9		0.0
Easy	136	72.8	128	58.9	67	41.8
Difficult		27.2		41.1		58.2
Mean		1.76		1.88		1.82
SD		0.20		0.17		0.18

Note: The DLBQ attitudes were originally designed to be rated on a 1-to-7 point semantic scale for each pair of attitudes, such that 1 was the negative end and 7 the positive end. In entering the questions into the online format in Survey Monkey, only the two endpoints were retained. Therefore, negative attitudes were scored a 1 and positive attitudes a 2; higher means indicate more positive attitudes, as they do in the original instrument, however, there is less variation in attitudes than would have been achieved had the original 7-point scale been maintained.

Determinants of intent to change behavior in each of the three domains (physical activity, diet and smoking) were ranked on a 5-point scale. A mean score for items in each domain was calculated, with a higher score indicating greater intent to change behavior toward a healthier lifestyle. The overall mean for intent to be more physically active was 3.07 (SD = 0.52), for eating healthy foods was 3.45 (SD = 0.61), and for stopping smoking was 3.57 (SD = .66).

Statistical Assumptions

Pearson's correlations and linear regression are the methods used in the data analysis. Pearson's correlations requires that the variables be at least interval level and have a linear relationship. All variables are interval (means of multiple items scored on

an ordinal level are interval variables). Scatter plots of all independent variables versus dependent variables revealed approximately linear relationships. Assumptions for correlation were met.

For all regression models, dependent variables are normally distributed based on P-P plot examination. Examination of residual plots indicated that residuals are distributed normally and that there are no problems with heteroskedasticity for any of the models. Variance Inflation Factors indicated no problems with multicollinearity for any of the models. Assumptions of linear regression were met for all models

Relationship between Quality of Life and Knowledge about Respiratory Disease

Research Question 1: To what extent does level of knowledge about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 1: There is an influence of the knowledge about LAM in women living with LAM on their quality of life.

Null Hypothesis 1: There is no influence of the knowledge about LAM in women living with LAM on their quality of life.

To answer the first research question, Pearson's correlation between quality of life measures (SGRQ) and knowledge scores on BCKQ were examined. The results are presented in Table 10. . Overall quality of life (total SGRQ) is not correlated to overall BCKQ score ($r = .229$, $p > .05$). Knowledge about breathlessness, inhaled bronchodilators, oral steroids and inhaled steroids are significantly related to total quality of life. That means that as knowledge in these areas increases, the overall SGRQ score increases. A higher SGRQ score indicates more symptoms and more adverse impacts;

that is, lower quality of life, hence higher knowledge is associated with lower quality of life.

Table 10

Bivariate Correlations between SGRQ and BCKQ

	SGRQ Total		SGRQ Symptoms		SGRQ Activity		SGRQ Impacts	
	r	*n*	*r*	*n*	*r*	*n*	*r*	*n*
BCKQ Total	0.229	65	.251[*]	66	0.185	96	0.03	93
Epidemiology	-0.063	63	-0.105	64	-0.072	92	-0.156	89
Etiology	0.114	63	0.125	64	0.055	92	-0.001	89
Symptoms	0.176	63	.322[**]	64	-0.017	92	0.011	89
Breathlessness	.336[**]	63	.363[**]	64	.310[**]	91	.223[*]	88
Phlegm	0.207	64	.271[*]	65	0.102	92	0.003	90
Infections	.356[**]	64	.390[**]	65	0.167	92	0.113	90
Exercise	0.004	63	0.039	64	-0.009	91	-0.145	88
Smoking	0.136	63	.282[*]	64	-0.049	90	0.041	88
Vaccination	0.106	64	0.192	65	0.107	95	0.083	92
Inhaled Bronchodilators	.462[***]	63	.500[***]	64	.361[***]	93	.261[*]	90
Antibiotics	0.219	63	.288[*]	64	0.136	91	0.081	88
Oral Steroids	.369[**]	63	.361[**]	64	.275[**]	90	0.183	88
Inhaled Steroids	.350[**]	63	.385[**]	64	.227[*]	92	0.104	90

*p < .05. **p < .01. ***p < .001.

SGRQ Symptoms scores are positively correlated with the following BCKQ knowledge areas: symptoms, breathlessness, phlegm, infections, smoking, inhaled bronchodilators, antibiotics, oral steroids, inhaled steroids, and overall knowledge. Again all the correlations are positive indicating that higher knowledge is associated with more severe symptoms.

SGRQ Activity scores are positively correlated with the following BCKQ knowledge areas: breathlessness, inhaled bronchodilators, oral steroids, and inhaled steroids. More knowledge in these areas is associated with more adverse effects of the LAM on activities of daily living.

SGRQ Impacts scores are positively correlated with the following BCKQ knowledge areas: breathlessness and inhaled bronchodilators. Greater knowledge in the areas of breathlessness and inhaled bronchodilators is associations with great impacts of the disease on respondents.

Correlations of less than .3 are considered small effects, .3 to .49 are considered medium effects, and .5 and above are considered large effects. Most of these significant correlations are in the medium range.

The null hypothesis of no relationship between knowledge and quality of life is rejected. There is a relationship between various aspects of quality of life and knowledge about some respondents relevant to respiratory disease. It appears that those who are more affected by LAM (that is, have more severe symptoms, more activity restrictions, and more impacts) are more knowledgeable with lower quality of life than those with fewer symptoms, less knowledgeable hence better quality of life.

Relationship of Quality of Life and Attitudes

Research Question 2: To what extent does the attitude about LAM in women living with LAM influence their quality of life?

Alternative Hypothesis 2: There is an influence of the attitude about LAM in women living with LAM on their quality of life.

Null Hypothesis 2: There is no influence of the attitude about LAM in women living with LAM on their quality of life.

To answer the second research question, bivariate correlations between attitudes about living with LAM, assessed by the BBQ, and quality of life were examined. The correlation matrix is presented in Table 11. Total quality of life is significantly negatively correlated with Confidence and positively correlated with Concerns and Disappointment. Quality of life is worse for those with less confidence in handling their disease, and those with more concerns about their disease and treatment and more disappointments in dealing with their disease. Having more severe symptoms is significantly positively correlated with more disappointments. Restricted activities because of LAM and impacts of the disease are both significantly correlated with all aspects of attitudes and behaviors except for nonadherence. Those with more restricted activities and greater impacts are less confident and satisfied with their treatment and have more concerns and disappointments. They do tend to have greater adherence to drug and treatment regimens than those with fewer activity restrictions and impacts (See Table 11). The null hypothesis of no relationship between quality of life and attitudes about treatment is rejected.

Table 11

Correlation between Quality of Life (SGRQ) and Attitude (BBQ)

		1	2	3	4	5	6	7	8	9	10
1 SGRQ Total	*R*	1	.855***	.938***	.957***	-.297*	.402**	-.234	.410***	.237	.141
	N	69	69	69	69	69	69	69	69	67	69
2 SGRQ Symptoms	*R*		1	.795***	.742***	-.208	.357**	-.166	.304*	.168	.063
	N		70	69	69	70	70	70	70	68	70
3 SGRQ Activity	*R*			1	.790***	-.235*	.293**	-.282**	.380***	.314**	.081
	N			104	100	104	104	104	104	102	104
4 SGRQ Impacts	*R*				1	-0.355***	.377***	-.301**	.502***	.273**	.167
	N				100	100	100	100	100	98	100
5 BBQ Confidence	*R*					1	-.230**	.673***	-.265**	.180*	-.414***
	N					143	143	143	143	140	142
6 BBQ Concerns	*R*						1	-.204*	.410***	.106	.161
	N						143	143	143	140	142
7 BBQ Satisfaction	*R*							1	-.254**	.024	-.220**
	N							143	143	140	142
8 BBQ Disappointment	*R*								1	.104	.341***
	N								143	140	142

(table continues)

		1	2	3	4	5	6	7	8	9	10
9 BBQ Adherence	*R*									1	-.292***
	n										140
10 BBQ Nonadherence	*R*										1
	N										142

*p < .05; **p < 01; ***p < .001.

Relationship between Quality of Life and Lifestyle Practices

Research Question 3: To what extent do the life style practices of women living with LAM influence their quality of life?

Alternative Hypothesis 3: There is an influence of life style practices of women living with LAM on their quality of life.

Null Hypothesis 3: There is no influence of life style practices of women living with LAM on their quality of life.

To answer the third research question, bivariate Pearson's correlations between quality of life and both attitudes toward lifestyle practices and determinants of behavioral change as measured by the DLBQ were examined. (Table 12)

Table 12

Correlations between Quality of Life (SGRQ) and Determinants of Lifestyle Behaviors (DLBQ)

		1	2	3	4	5	6	7	8	9	10
1 SGRQ Total	r	1	.855***	.938***	.957***	-.479***	-.074	-.057	-.494***	-.125	.062
	n	69	69	69	69	69	69	53	69	69	56
2 SGRQ Symptoms	r		1	.795***	.742***	-.410***	-.114	.078	-.448***	-.202	-.074
	n		70	69	69	70	70	54	70	70	57
3 SGRQ Activity	r			1	.790***	-.457***	-.213*	-.038	-.435***	-.157	.153
	n			104	100	104	104	59	104	104	64
4 SGRQ Impacts	r				1	-.482***	-.104	-.143	-.521***	-.131	.008
	n				100	100	100	59	100	100	64
5 DLBQ Physical Activity	r					1	.219*	.161	.386***	.042	.052
	n					136	129	64	136	129	69
6 DLBQ Healthy Food	r						1	.069	.148	.395***	.107
	n						129	64	129	129	69
7 DLBQ Smoking	r							1	-.107	.168	.294*
	n							64	64	64	64

(table continues)

		1	2	3	4	5	6	7	8	9	10
8 DLBQ Attitudes Physical Activity	r								1	.277**	.174
	n								136	129	69
9 DLBQ Attitudes Healthy Food	r									1	.038
	n									129	69
7 DLBQ Attitudes Smoking	r										1
	n										69

*p < .05; **p < .01; ***p < .001.

The total SGRQ quality of life scores and all subscale scores were significantly negatively correlated to the physical activity attitudes and intent to change of the DLBQ. As quality of life decreases (i.e., SGRQ increases), intent to be more physically active is lower (lower scores on DLBQ indicate less intent to change) and attitudes toward physical activity are more negative. There were no significant relationships between quality of life and attitudes or intent to change diet or smoking behaviors.

The null hypothesis of no relationship between quality of life and lifestyle practices can be rejected. Those with more positive attitudes toward physical activity and greater intent to become more physically active, have a higher quality of life

Predicting Quality of Life from KAPs

Linear regression was used to examine the relationship of quality of life (total and symptoms, activity and impacts) with KAPs, controlling for household income and education. Only variables that were significantly correlated with the outcome variable were included in each model along with income and education as a controls being entered in a first step followed by the addition of the other variables. Because of strong correlation between attitudes and intent to chance scores of DLBQ for physical activity, only intent to change was included in the regression models. Based on univariate and correlation analyses, the following regression models were tested.

$$
\begin{aligned}
SGRQ\ Total = \ & b_0 + b_1 Income\ \$70k \\
& + b_2 Associates\ Degree + b_3 Bachelors\ Degree \\
& + b_4 Graduate\ Degree + b_5 BBQ\ Confidence + b_6 BBQ\ Concerns \\
& + b_7 BBQ\ Disappointment + b_8 BCKQ\ Breathlessness \\
& + b_9 BCKQ\ Infections + b_{10} BCKQ\ Inhaled\ Bronchodilators \\
& + b_{11} BCKQ\ Oral\ Steroids + b_{12} BCKQ\ Inhaled\ Steroids \\
& + b_{13} DLBQ\ Physical\ Activity
\end{aligned}
$$

To reduce the number of independent variables because of small sample size[1], a regression model using only total knowledge score instead of individual knowledge scores was also tested. (Rule of thumb in regression is 1 independent variable for each 10 cases. There are approximately 60 cases with all variables for regression, suggesting a maximum of 6 independent variables would be appropriate. It turned out that the model with more independent variables was a better fit, however.)

$$SGRQ\ Total = \ b_0 + b_1 Income\ \$70k$$
$$+ b_2 Associates\ Degree + b_3 Bachelors\ Degree$$
$$+ b_4 Graduate\ Degree + b_5 BBQ\ Confidence + b_6 BBQ\ Concerns$$
$$+ b_7 BBQ\ Disappointment + b_8 BCKQ\ Total$$
$$+ b_9 DLBQ\ Physical\ Activity$$

The results of both of these models predicting overall quality of life are presented in Table 12. When entered into the equation with education, income was a significant predictor of quality of life. Having an associate's degree significantly predicated higher quality of life than having only high school degree or less. Once beliefs and behaviors knowledge, and determinants of change were added, income was no longer significant, although having an associate's degree remained significant. The model with individual knowledge subscale scores (Model 2 in Table 13) explains more of the variance (58%) than the model with total knowledge score (Model 4; 43%) so is a better fitting model. The only significant predictor of total quality of life, after controlling for income and education, was DLBQ Activity. A 1- point increase in determinants of physical activity change resulted in a 9.14 drop in SGRQ score, indicating that the more likely one is to increase physical activity the better the total quality of life.

Table 13

Predictors of Quality of Life (SGRQ Total)

| | Total Quality of Life | | | | | |
| | Model 1 | Model 2 | | Model 3 | Model 4 | |
Variable	*B*	*B*	95% CI	*B*	*B*	95% CI
Constant	28.72***	-2.86	[-69.74, 64.01]	28.48***	2.73	[-66.59, 72.05]
Income < $70k	16.60**	9.08	[-.49, 18.65]	17.02**	9.40	[-.65, 19.46]
Associate degree	21.68*	16.78*	[1.93, 31.64]	21.77*	14.12	[-2.23, 30.48]
Bachelors degree	6.27	3.52	[-7.52, 14.55]	6.75	4.35	[-7.69, 16.40]
Graduate degree	11.09	4.91	[-7.93, 17.75]	12.16	8.58	[-4.79, 21.94]
BBQ Confidence		-.24	[-7.06, 6.58]		.78	[-6.26, 7.82]
BBQ Concerns		9.95	[-3.18, 23.07]		10.39	[-2.92, 23.70]
BBQ Disappointment		4.25	[-2.43, 10.94]		5.38	[-1.72, 12.49]
BCKQ Breathlessness		12.03	[-5.95, 30.02]			
BCKQ Infections		14.94	[-1.56, 31.44]			
BCKQ Inhaled Bronchodilators		9.34	[-10.26, 28.94]			
BCKQ Oral Steroids		-3.34	[-20.22, 13.53]			
BCKQ Inhaled Steroids		4.78	[-17.70, 27.27]			
DLBQ Physical Activity		-9.14*	[-18.27, -.10]		-0.57*	[-19.73, -1.42]
BCKQ Total					16.40	[-4.25 37.06]
R^2	.21	.58		.22	.43	
F	3.84**	5.15***		4.21**	4.60***	
ΔR^2		.37***			.21**	
ΔF		1.31			.39	
N	62	62		64	64	

*p < .05. **p < .01. ***p < .001

Predicting Quality of Life Subscales

Regression models were run for each of the SGRQ subscales with independent variables selected if the bivariate correlations were significant. Again income and education were entered as controls. In the model predicting SGRQ Activity, age was also added as a control because one-way ANOVA (see above) had indicated that there was a significant difference in Activity score by age group. The three regression models are as follows:

$$
\begin{aligned}
SGRQ\ Symptoms \\
= b_0 + b_1 Income\ under\ \$70k \\
+ b_2 Associates\ Degree + b_3 Bachelors\ Degree \\
+ b_4 Graduate\ Degree + b_5 45 - 54\ Years + b_6 55\ and\ older \\
+ b_7 BBQ\ Disappointment + b_8 BCKQ\ Total \\
+ b_9 DLBQ\ Physical\ Activity
\end{aligned}
$$

$$
\begin{aligned}
SGRQ\ Activity \\
= b_0 + b_1 Income\ under\ \$70k \\
+ b_2 Associates\ Degree + b_3 Bachelors\ Degree \\
+ b_4 Graduate\ Degree + b_5 45 - 54\ Years + b_6 55\ and\ older \\
+ b_7 BBQ\ Confidence + b_8 BBQ\ Concerns + b_9 BBQ\ Satisfaction \\
+ b_{10} BBQ\ Disappointments + b_{11} BBQ\ Adherence \\
+ b_{12} BCKQ\ Breathlessness + b_{13} BCKQ\ Inhaled\ Bronchodilators \\
+ b_{14} BCKQ\ Oral\ Steroids + b_{15} BCKQ\ Inhaled\ Steroids \\
+ b_{16} DLBQ\ Physical\ Activity
\end{aligned}
$$

$$
\begin{aligned}
SGRQ\ Impacts \\
= b_0 + b_1 Income\ under\ \$70k \\
+ b_2 Associates\ Degree + b_3 Bachelors\ Degree \\
+ b_4 Graduate\ Degree + b_5 45 - 54\ Years + b_6 55\ and\ older \\
+ b_7 BBQ\ Confidence + b_8 BBQ\ Concerns + b_9 BBQ\ Satisfaction \\
+ b_{10} BBQ\ Disappointments + b_{11} BBQ\ Adherence \\
+ b_{12} BCKQ\ Breathlessness + b_{13} BCKQ\ Inhaled\ Bronchodilators \\
+ b_{14} DLBQ\ Physical\ Activity
\end{aligned}
$$

The results of these three models are presented in Table 14. Intent to increase physical activity is the only significant predictor of symptoms quality of life after controlling for income, education, and age. The significant predictor of activity component of quality of life was intended to increase physical activity after controlling for income, education, and age. Note that age also significantly predicted activity quality of life, with those 45-to-54 and 55 and over both having significantly higher scores (indicating more effects of the disease on activity) than those younger than 45. Satisfaction was marginally significant predictors of activity component of quality of life. Impacts component of quality of life was significantly predicted by disappointments

and intent to increase physical activity. Adherence was a marginally significant predictor

of impacts quality of life.

Table 14

Predictors of Quality of Life Symptoms, Activity, and Impacts

	Symptoms		Activity		Impacts	
Variable	B	95% CI	B	95% CI	B	95% CI
Constant	52.47*	[4.05, 10.78]	34.47	[-39.21, 108.141]	5.40	[-49.27, 60.06]
Income < $70k	11.27†	[-.95, 23.49]	8.77†	[-1.13, 18.67]	4.58	[-3.03, 12.19]
Associate's degree	19.75†	[-.72. 40.22]	11.25	[-6.17, 28.66]	3.46	[-9.70, 16.61]
Bachelor's degree	5.97	[-8.95, 20.90]	7.97	[-5.53, 21.47]	-.92	[-11.34, 9.51]
Graduate degree	6.34	[-10.29, 22.97]	6.67	[-8.37, 21.71]	-.81	[-12.21, 10.59]
45-to-54 year olds			14.23*	[3.01, 25.44]		
55 and older			15.41*	[2.97, 27.85]		
BBQ Confidence			1.41	[-9.18, 11.99]	-4.65	[-12.78, 3.46]
BBQ Concerns			10.93	[-2.75, 24.60]	6.37	[-3.97, 16.71]
BBQ Satisfaction			-8.01†	[-17.02, 1.00]	-.98	[-7.83, 5.87]
BBQ Disappointment	3.70	[-4.57, 11.98]	2.18	[-5.19, 9.55]	8.39**	[2.61, 14.17]
BBQ Adherence			5.35	[-1.81, 12.52]	4.96†	[-0.33, 10.25]
BCKQ Breathlessness			15.21	[-5.61, 36.03]	4.72	[-11.04, 20.49]
BCKQ Inhaled Bronchodilators			6.88	[-13.93, 27.68]	8.48	[-4.03, 20.981]
BCKQ Oral Steroids			-3.88	[-21.21, 13.46]		
BCKQ Inhaled Steroids			17.55	[-7.80, 42.69]		
DLBQ Physical Activity	-13.29*	[-24.58, -2.01]	-15.12**	[-25.53, -4.71]	-8.45*	[-16.38, -.53]
BCKQ Total	26.07*	[.18, 51.95]				
R^2	.32		.55		.50	
F	3.82**		5.00***		5.63***	
N	65		83		82	

†$p < .10$. *$p < .05$. **$p < .01$. ***$p < .001$.

The null hypothesis of no relationship between quality of life and all the

independent variables (knowledge, attitudes, and lifestyle practices) can be rejected.

Only intent to increase physical activity remains a consistently significant predictor of

overall quality of life after controlling for income, education, knowledge, and attitudes.

Summary

The results of the data analysis for this study rejected all three null hypotheses; showing that there is relationship between KAPs and the quality of life. Descriptive statistic described the sample population and the collected data. Pearson's correlation and linear regression were the methods used in data analysis and utilized for each one of the independent variables (KAPs), in order to examine each variable's relation to the quality of life.

To answer the first research question, Pearson's correlation between quality of life measures (SGRQ) and knowledge scores on BCKQ were examined. The null hypothesis of no relationship between knowledge and quality of life was rejected, determining that there is a relationship between various aspects of quality of life and knowledge about some subjects relevant to respiratory disease. Knowledge about breathlessness, inhaled bronchodilators, oral steroids, and inhaled steroids were significantly related to total quality of life. It appeared that those with lower quality of life (that is, have more severe symptoms, more activity restrictions, and more impacts of LAM) are more knowledgeable than those with better quality of life

To answer the second research question, bivariate correlations between attitudes about living with LAM assessed by the BBQ, and quality of life were examined and the null hypothesis of no relationship between quality of life and attitudes about treatment was rejected showing total quality of life is significantly negatively correlated with Confidence and positively correlated with Concerns and Disappointment. Quality of life is worse for those with less confidence in handling their disease, those with more

concerns about their disease and treatment, and those with more disappointments in dealing with their disease.

To answer the third research question, bivariate Pearson's correlations between quality of life and both attitudes toward lifestyle practices and determinants of behavioral change as measured by the DLBQ were examined. The null hypothesis of no relationship between quality of life and lifestyle practices was rejected. Those with more positive attitudes toward physical activity and greater intent to become more physically active, had a higher quality of life.

Implications for Social Change

The desired outcome achieved by this study is crucial to social change, because it encourages researchers to initiate more studies on KAPs within LAM population; in particular future research on the KAPs of the TSC LAM population since the focus of the present study was women with sporadic LAM. Through the awareness brought on by the results of this study the stakeholders are encouraged to promote initiatives that would improve knowledge, attitude, and healthier life style practices. These initiatives could enhance and create much needed hope and encouragement to possibly enhance quality of life. It is apparent that the findings of the present study contribute to positive social change at all levels.

Conclusion

LAM is a rare cystic multisystem lung disease which almost exclusively affects women of childbearing age (King 2010). There is no cure for LAM and the treatment for women with this debilitating disease is mainly supportive (Cottin et al., 2011). It is

essential to be adamant about the importance of initiatives to promote patient's knowledge about LAM, patient's attitude about LAM, and lifestyle practices that positively impact quality of life of women with LAM. Knowledge can be essential not only for the health care practitioners but also for the patients to understand how well they could manage living with LAM. Attitude and healthy lifestyle practices could also allow these targeted populations to improve the overall quality of their life. LAM is a devastating disease that eventually takes away the ability of patients to be productive and have quality of life. The importance of improved KAPs and its relation to the quality of life have been proven to be effective with other chronic and respiratory diseases. The fact that there is no definitive treatment at the present time for LAM, makes it imperative to take effective measures at all levels to promote and initiate studies as well as programs that would improve knowledge with clarity in regards to LAM, promoting effective attitude, and healthier lifestyle practices within the LAM community.

The following Chapter 5 will analyze and interpret the key findings. This chapter will also discuss the social change implications of the present study findings as well as describing the recommendations for further research, and initiatives improving KAPs, and related quality of life for LAM community.

Chapter 5: Discussion, Conclusions, and Recommendations

Introduction

The purpose of this study was to examine the effects of KAPs of the sporadic

LAM patients registered at the LAM Foundation and how these variables related to their

quality of life. Although there have been many efforts by the researchers to look at the

etiology of LAM there has been a noticeable gap in the area of LAM patients KAPs and

quality of life. The aim of conducting this study was to encourage more focus and

initiatives to improve the KAPs within the LAM community at large.

Interpretation of the Findings

This research study utilized the quantitative study design incorporating the survey

research strategy in order to examine the KAPs of the LAM population and their effect

on the quality of life. Collected data included the descriptive and demographic

characteristics of the study population. There were no available data on the population of

sporadic LAM patients and the only information available was from the NHLBI's LAM

registry (Ryu et al., 2006), which provided age at time of enrollment in the registry as

well as race. The current sample were significantly older (mean age 47.7 years) than the

age of NHLBI's LAM enrollees (45.1±.69), $z = 3.768$, $p < .001$. This study population

also had a higher percentage of Whites (92.5%) than the NHLBI sporadic LAM enrollees

(87.2%). It should be noted, however, that there is no indication that the NHLBI registry

is representative of the sporadic LAM population; it is simply the only demographic

information publically available. The BCKQ, the DLBQ, the BBQ, and the SGRQ

instrumentation were employed as tools measuring the knowledge, attitude, practices, as

well as quality of life. Pearson correlation and linear regression showed the relation between each independent variable (KAPs) to the dependent variable (quality of life). TTM, focuses on decision-making of the target population and their intentions to change behavior, supported the study's expectations that the depended variable (quality of life) was influenced by the independent variables (KAPs).

The findings of statistical analysis in this study showed there was a relationship among the KAPs and the quality of life of the sporadic LAM patients. Although there have been no head to head studies to compare the findings of the present study; it is quite reasonable to say that these results were complementary to the previously mentioned studies related to the KAPs of the other chronic and respiratory diseases

Based on the findings knowledge about the breathlessness, inhaled bronchodilators, and oral steroids were significantly related to the quality of life. It appeared that those who were more affected by LAM having more severe symptoms, more activity restrictions, and more impacts were more knowledgeable about their disease than those who were not as affected by their disease symptoms. The assumption for this correlation would suggest that greater knowledge in these areas could have been brought on by patients having had longer duration of disease. This could have encouraged them to seek more knowledge about their disease, and therefore become more aware of the symptoms, and adverse impacts which associated with lower quality of life. Although this finding was contradictory to previous studies showing improved quality of life with more knowledge, the results showed a significant relationship between knowledge and quality of life. The interpretation could lead to this understanding that longer duration of

disease would expose the patients to learning, and knowing more about the condition, and symptoms that could cause concerns and restriction on quality of their life. The interpretation of this finding call for specific and perhaps a higher level of LAM knowledge questionnaire to examine the relationship knowledge has on quality of life. Patients have to confront their illness and becoming knowledgeable about the facts and myths of LAM. This could allow women to change behavior and lifestyle modifications in order to possibly improve their quality of life.

Based on the findings, the higher qualities of life were observed with those who scored higher with confidence in handling their disease and were more positive about dealing with concerns. This result was consistent with Holman and Lorig's (2004) findings that the patient's quality of life and survival were much better if the patient's had a positive attitude and strived to overcome the concerns caused by their chronic condition. The present study results also showed that those who were less restricted with activities and had less impact were more confident and less concerned and disappointed. These patients were more willing and had more adherences to drug and treatment. LAM is a debilitating disease that can potentially suffocate women (LAM Foundation, 2013); promoting positive attitude can be conducive to LAM patients understanding and managing their chronic disease. As stated by Haq et al. (2013) in their study findings the way patients behaved towards their disease condition was dependent on the attitude they had and their behavior was very much depended on the attitude they held. Therefore, Haq et al. (2013) concluded that positive attitude will encourage positive behavior, which ultimately brings about healthier practices by the patients. Mancuso et al. (2010),

elaborated on their study findings by stating correlation between knowledge and attitude as well as knowledge and self-efficacy were lower than correlation between attitude and self-efficacy among the study population which in turn negatively affected the quality of life of asthma patients.

The present study results with lifestyle practices and how they related to the quality of life showed those who had more positive attitudes towards physical activity had a higher quality of life. This study implied that those who had higher intent to change behavior towards healthier food and physical activity had a higher quality of life. The importance of healthier lifestyle practices has been emphasized with chronic diseases, in particular physical activity. This study's results were consistent with Yang and Chen's (2005) findings that maintaining physical activity will slow down the deterioration of pulmonary function, will allow for daily activities to maintain, and ultimately enhance quality of life.

Limitations of the Study

This research was limited to the sporadic LAM population who were registered with the LAM Foundation, because it holds the largest number of LAM patients to date (LAM Foundation, 2014). Pulling samples from one registry limited access to all LAM patients, therefore limited generalizability of the results to all patients. Another limitation was reduced sample available for some analysis. Based on complaints from the first set of respondents, a change in how the online survey worked was made. Originally all questions had to be answered. A change was made to make all questions optional except the consent .This limitation reduced the sample available for some analyses, therefore

reducing internal validity. It should be noted that the response rate was only 13.8% of those who were eligible to participate which they did; however, this may not be representative of all the LAM population. Although this response rate was less than 14%, according to Hamilton (2009) 13.35% is the overall response rate for online surveys. The other limitation is the fact that the study population was limited to only sporadic LAM patients and not TSC LAM patients; that restricted the findings to only serve the sporadic LAM patients and not TSC LAM patients. The study population had good scores in knowledge of respiratory disease, and the assumption of the study results considers further limitation that LAM registry population could have been more educated in regards to LAM than other LAM patients elsewhere, and this might have been a cause for bias. The fact that I am a LAM patient myself could also be another reason for bias; however, this research was conducted solely based on an objective and scholarly manner. Additionally, because the study questionnaires were not specifically designed for LAM, it may have caused bias and limitation to this study.

Recommendations

The recommendations for future research should definitely extend to further investigations, and focus in KAPs studies for all LAM patients, because the population in the present study represented only sporadic LAM patients. It was evident from the present findings that there are strong correlations between KAPs and quality of life. Although the correlation between knowledge of this study population and quality of life was contradictory to the previous studies indicating the more knowledge of the disease the better quality of life; the future studies should further investigate the relationship

between the knowledge in particular LAM knowledge and its relation to quality of life. The findings of the present study encourage the need for future efforts, and initiatives to develop specific KAPs questionnaires for LAM to measure these variables and their relation to quality of life.

Conclusion

LAM patients are living with disease that is rare, prograssive, and most often fatal (King, 2010). The findings of this study implied that there are significant correlations among KAPs and quality of life within the study population . It certainly encourages further investigations in regards to the specific LAM KAPs and quality of life among all LAM population. The improved quality of life due to the positive attitudes and lifestyle practices of this study population encourages LAM community as well as other stake holders to implement programs, workshops, and interventions that could promote such findings further among all LAM community.

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Appendix A: Agreement letter from LAM Foundation

November 22, 2013

Dear Ms Vafamand,

Thank you for sharing your dissertation proposal with me. I am happy to assist you in reaching out to LAM patients. As we discussed, I will send an email to each LAM patient registered with the Foundation in order to collect your data as noted in your proposal.

If I can be of further assistance, please let me know. Good luck!

Sincerely
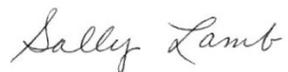
Sally Lamb

Sally Lamb
Director of Patient Services
The LAM Foundation

Appendix B: Permission to use BCKQ

Subject: Fw: permission

From: Shahpar Vafamand (shahparvafamand@yahoo.com)

To: peacedall5.sv@gmail.com;

Date: Monday, December 9, 2013 10:36 AM

On , Shahpar Vafamand <shahparvafamand@yahoo.com> wrote:

On Thursday, December 5, 2013 2:02 PM, Roger White <roger.white4@virgin.net> wrote:
I am pleased to send you a copy.
Regards
Roger White

From: Shahpar Vafamand [mailto:shahparvafamand@yahoo.com]
Sent: 05 December 2013 18:39
To: roger.white4@virgin.net
Subject: permission

Hello Dr White,

My name is Shahpar Vafamand and I am a doctoral student in Public Health at Walden University. I am pursuing my dissertation in a rare lung disease called Lymphangioleiomyomatosis (LAM) and focusing on the area of Knowledge, attitude, and life style practices of these targeted population. I have been looking for a questionnaire that would be related to the KAPs of patients with chronic lung disease since there are none put together for LAM in particular. I have not had any luck although I came across your valuable tool the BCKQ and I was extremely excited. I would be very grateful if you could please assist with obtaining a copy of this questionnaire as well the scoring procedure and most of all permission to use it in my study. I would also welcome any of your expert recommendation or suggestion if you know of questionnaire that would address not only knowledge but also attitude and lifestyle practices of these target population. I thank you so much for your time and assistance.

sincerely yours,

Shahpar Vafamand

Appendix C. Permission to use SGRQ

St George's
University of London

Medicine, Biomedical Sciences, Health and Social Care Sciences

19 August 2013

Cranmer Terrace
London SW17 0RE

Switchboard
+44 (0)20 8672 9944

www.sgul.ac.uk

To Whom It May Concern:

This is to confirm that St George's University of London (St George's Hospital Medical School) has given permission for Shahpar Vafamand, doctoral student in public health at Walden University, USA to use the SGRQ in a study looking at the quality of life in patients with Lymphangioleiomyomatosis (LAM).

Professor Paul Jones, PhD FRCP
Professor of Respiratory Medicine

P.W. Jones, PhD FRCP
Professor of Respiratory Medicine

Appendix D: BCKQ

NHS BRISTOL COPD KNOWLEDGE QUESTIONNAIRE (BCKQ)©

Name: _____ **Date:** _____

This questionnaire is designed to find out what you know about your lung problem. It should be completed without help from anyone else. This usually takes between 10 and 20 minutes. Your answers will help us to find out what information you need to help you to understand and manage your lung condition.

Mark the circle which you think is the correct answer.

1 In COPD:

		True	False	Don't Know
a	In COPD the word "chronic" means it is severe.	○	○	○
b	COPD can only be confirmed by breathing tests.	○	○	○
c	In COPD there is usually gradual worsening over time.	○	○	○
d	In COPD oxygen levels in the blood are always low.	○	○	○
e	COPD is unusual in people less than 40 years old.	○	○	○

2 COPD:

		True	False	Don't Know
a	More than 80% of COPD cases are caused by cigarette smoking.	○	○	○
b	COPD can be caused by occupational dust exposure.	○	○	○
c	Longstanding asthma can develop into COPD	○	○	○
d	COPD is commonly an inherited disease.	○	○	○
e	Women are less vulnerable to the effects of cigarette smoking than men.	○	○	○

3 The following symptoms are *COMMON* in COPD:

		True	False	Don't Know
a	Swelling of ankles	○	○	○
b	Fatigue (tiredness)	○	○	○
c	Wheezing	○	○	○
d	Crushing chest pain	○	○	○
e	Rapid weight loss	○	○	○

4 Breathlessness in COPD:

		True	False	Don't Know
a	Severe breathlessness prevents travel by air.	○	○	○
b	Breathlessness can be worsened by eating large meals.	○	○	○
c	Breathlessness means that your oxygen levels are low.	○	○	○
d	Breathlessness is a normal response to exercise.	○	○	○
e	Breathlessness is primarily caused by a narrowing of the bronchial tubes.	○	○	○

5 Phlegm (sputum):

		True	False	Don't Know
a	Coughing phlegm is a common symptom in COPD	○	○	○
b	Clearing phlegm is more difficult if you get dehydrated.	○	○	○
c	Bronchodilator inhalers can help clear phlegm.	○	○	○
d	Phlegm causes harm if swallowed.	○	○	○
e	Clearing phlegm can be assisted by breathing exercises.	○	○	○

6 Chest infections / exacerbations:

		True	False	Don't Know
a	Chest infections often cause coughing of blood.	○	○	○
b	With chest infections phlegm usually becomes coloured (yellow or green).	○	○	○
c	Exacerbations (episodes of worsening) can occur in the absence of a chest infection.	○	○	○
d	Chest infections are always accompanied by a high temperature.	○	○	○
e	Steroid tablets should be taken whenever there is an exacerbation.	○	○	○

7 Exercise in COPD:

		True	False	Don't Know
a	Walking is better exercise than breathing exercises to improve fitness.	○	○	○
b	Exercise should be avoided as it strains the lungs.	○	○	○
c	Exercise can help maintain your bone density.	○	○	○
d	Exercise helps relieve depression.	○	○	○
e	Exercise should be stopped if it makes you breathless.	○	○	○

8 Smoking:

		True	False	Don't Know
a	Stopping smoking will reduce the risk of heart disease.	○	○	○
b	Stopping smoking will slow down further lung damage.	○	○	○
c	Stopping smoking is pointless as the damage is done.	○	○	○
d	Stopping smoking usually results in improved lung function.	○	○	○
e	Nicotine replacement therapy is only available on prescription.	○	○	○

9 Vaccination:

		True	False	Don't Know
a	A flu jab is recommended every year.	○	○	○
b	You can get flu from having a flu jab.	○	○	○
c	You can only have a flu jab if you are 65 or over.	○	○	○
d	A pneumonia jab protects against all forms of pneumonia.	○	○	○
e	You can have a pneumonia jab and a flu jab on the same day.	○	○	○

10 Inhaled brochodilators:

		True	False	Don't Know
a	All bronchodilators act quickly (within 10 minutes).	○	○	○
b	Both short and long acting bronchodilators can be taken on the same day.	○	○	○
c	Spacers (e.g. volumatic, nebuhaler, aerochamber) should be dried with a towel after washing.	○	○	○
d	Using a spacer device will increase the amount of drug deposited in the lungs.	○	○	○
e	Tremor may be a side effect of bronchodilators.	○	○	○

11 Antibiotic treatment in COPD:

		True	False	Don't Know
a	To be effective, the course should last at least 10 days.	○	○	○
b	Excessive use of antibiotics can cause resistant bacteria (germs).	○	○	○
c	Antibiotics will clear all chest infections.	○	○	○
d	Antibiotic treatment is necessary for an exacerbation (worsening) however mild.	○	○	○
e	You should seek advice if antibiotics cause severe diarrhoea.	○	○	○

12 Steroid tablets given for COPD (eg Prednisolone):

		True	False	Don't Know
a	Steroid tablets help strengthen muscles.	○	○	○
b	Steroid tablets should be avoided if there is a chest infection.	○	○	○
c	The risk of long-term side effects due to steroids is less with short courses than with continuous treatment.	○	○	○
d	Indigestion is a common side effect from using steroid tablets.	○	○	○
e	Steroid tablets can increase your appetite.	○	○	○

13 Inhaled steroids (brown, red or orange):

		True	False	Don't Know
a	Inhaled steroids should be stopped if you are given steroid tablets.	○	○	○
b	Steroid inhalers can be used for rapid relief of breathlessness.	○	○	○
c	Spacer devices reduce the risk of getting thrush in the mouth.	○	○	○
d	Steroid inhaler should be taken before your bronchodilator.	○	○	○
e	Inhaled steroids improve lung function in COPD.	○	○	○

Further supplies can be obtained from:
Department of Medicine (BCKQ)
Frenchay hospital,
Bristol BS16 1LE

Ref.: White R, Walker P, Roberts S, Kalisky S, White P,
Chronic Respiratory Disease. 2006;3:123-131

Designed by the Medical Illustration Department, Frenchay Hospital, Bristol

Appendix E: DLBQ

Determinants of Lifestyle Behavior Questionnaire
DLBQ

Items

Item no.	Items and answering categories	PHYSICAL ACTIVITY		TPB Construct
1	In my opinion, being	unpleasant _ _ _ _	pleasant	**Attitude**
2	more physically active	frustrating _ _ _ _	satisfactory	Affective
3	is . . .	bad _ _ _ _	good	**Attitude**
4		unimportant _ _ _ _	important	Cognitive
5		desirable _ _ _ _	desireable	
6		difficult _ _ _ _	easy	
7	My partner thinks that I should be more physically active	Totally disagree;		Subjective norms
8	My family thinks that I should be more physically active	partially disagree; neither agree nor disagree;		
9	My friends think that I should be more physically active	partially agree; totally agree		
10	I am able to be more physically active under normal circumstances			PBC Perceived control
11	I am able to be more physically active, even when I am busy or on holidays			
12	I find it difficult to be more physically active under normal circumstances			PBC Perceived difficulty
13	I find it difficult to be more physically active when I am busy			
14	When I have not been able to be physically active for a while (e.g., because of illness) it is hard for me to start again			
15	I intend to be more physically active within two months			Intention

Determinants of Lifestyle Behavior Questionnaire
DLBQ

Items

Item no.	Items and answering categories	DIETARY BEHAVIOR		TPB Construct
1	In my opinion, eating	unpleasant _ _ _ _ _ pleasant		Attitude Affective
2	healthier food is . . .	frustrating _ _ _ _ _ satisfactory		
3	(Eating healthier food	bad _ _ _ _ _ good		Attitude Cognitive
4	means: a diet with less	unimportant _ _ _ _ _ important		
5	(saturated) fats, more	desirable _ _ _ _ _ desireable		
6	vegetables and fruit, and less calories)	difficult _ _ _ _ _ easy		
7	My partner thinks that I should eat healthier food	Totally disagree;		Subjective norms
8	My family thinks that I must eat healthier food	partially disagree; neither agree nor disagree;		
9	My friends think that I must eat healthier food	partially agree;		
10*	I am able to eat healthy food under normal circumstances	totally agree		PBC Perceived control
11	I am able to eat healthy food in an environment in which a lot of unhealthy food is offered (e.g. a canteen or company restaurant)			
12	I am able to eat healthy food, even when others offer me less healthy food (e.g., at birthday parties or other parties)			
13	I am able to eat healthy food, even when I am busy			
14	I find it difficult to eat healthy food under normal circumstances			PBC Perceived difficulty

Determinants of Lifestyle Behavior Questionnaire
DLBQ

Items

Item no.	Items and answering categories	DIETARY BEHAVIOR	TPB Construct
15	I find it difficult to eat healthy food in an environment in which a lot of unhealthy food is offered	Totally disagree; partially disagree; neither agree nor disagree; partially agree; totally agree	PBC Perceived difficulty (continued)
16	I find it difficult to eat healthy food when I am busy		
17	I find it difficult to eat healthy food when others offer me less healthy food		
18	I find healthy food too expensive		Perceived barriers
19*	It takes too much time to eat healthy food every day		
20	I intend to eat healthier food within two months		Intention
21	I think that I will eat healthier within two months		

Note. TPB = theory of planned behavior; PBC = perceived behavioral control.
*This item was not retained in the final model.

Determinants of Lifestyle Behavior Questionnaire
DLBQ

Items

Item no.	Items and answering categories		SMOKING BEHAVIOR	TPB Construct
1	In my opinion, to stop	unpleasant _ _ _ _ _ pleasant		**Attitude**
2	smoking is . . .	frustrating _ _ _ _ _ satisfactory		Affective
3		bad _ _ _ _ _ good		**Attitude**
4		unimportant _ _ _ _ _ important		Cognitive
5		desirable _ _ _ _ _ desireable		
6*		difficult _ _ _ _ _ easy		
7	My partner thinks that I should stop smoking	Totally disagree;		**Subjective norms**
8	My family thinks that I should stop smoking	partially disagree;		
9	My friends think that I should stop smoking	neither agree nor disagree;		
10*	I am able to smoke less under normal circumstances	partially agree;		**PBC** Perceived control
11	I am able to stop smoking under normal circumstances	totally agree		
12	I am able to refrain from smoking even when others offer me a cigarette/cigar			
13	I am able to refrain from smoking when others around me are smoking			
14	I am able to refrain from smoking under stressful circumstances			
15*	I find it difficult to smoke less under normal circumstances			**PBC** Perceived difficulty
16	I find it difficult not to smoke when others around me are smoking			

Determinants of Lifestyle Behavior Questionnaire
DLBQ

Items

Item no.	Items and answering categories	SMOKING BEHAVIOR	TPB Construct
17	I find it difficult not to smoke when others insist or offer me a cigarette	Totally disagree; partially disagree;	PBC Perceived difficulty (continued)
18	I find it difficult not to smoke under stressful circumstances	neither agree nor disagree;	
19*	I think that I will stop smoking within the next two months	partially agree; totally agree	
20	I intend to stop smoking within a year		Intention
21	I do not intend to stop smoking		

Note. TPB = theory of planned behavior; PBC = perceived behavioral control.
*This item was not retained in the final model.

Appendix F: BBQ

◣ PsycTESTS®

**Beliefs and Behaviour Questionnaire
BBQ**

Items
_____

1. I have sufficient understanding about my illness.
2. I have sufficient understanding about the options for managing my illness.
3. I know what to expect from my illness management.
4. The management of my illness is a mystery for me.
5. Using any medication involves some risk.
6. Natural remedies are safer than medicines.
7. The management of my illness disrupts my life.
8. My doctors are very knowledgeable.
9. My current management will keep my illness at bay.
10. I am receiving the best possible management.

Demographic information collected
11. Financial difficulties limit my access to the best healthcare.
12. I am on too many medications.
13. I am concerned about the side effects from my medications.
14. I am satisfied with the information my doctors share with me.
15. My doctors have limited management options to offer me.
16. My doctors spend adequate time with me.
17. My doctors are compassionate.
18. I have a say in the way my illness is managed.
19. My medications are working.
20. It is helpful to know the experiences of others with similar illness as mine.
21. It is unpleasant to use some of my medications.
22. It is physically difficult to handle some of my medications.
23. I vary my recommended management based on how I am feeling.
24. I put up with my medical problems before taking any action.
25. I push myself to follow the instructions of my doctors.
26. I get confused about my medications.
27. I make changes in the recommended management to suit my lifestyle.
28. I ensure I have enough medications so that I do not run out.
29. I have strict routines for using my regular medications.
30. I keep my medications close to where I need to use them.

Beliefs and Behaviour Questionnaire
BBQ

Items

1. I have sufficient understanding about my illness.
2. I have sufficient understanding about the options for managing my illness.
3. I know what to expect from my illness management.
4. The management of my illness is a mystery for me.
5. Using any medication involves some risk.
6. Natural remedies are safer than medicines.
7. The management of my illness disrupts my life.
8. My doctors are very knowledgeable.
9. My current management will keep my illness at bay.
10. I am receiving the best possible management.

Demographic information collected

11. Financial difficulties limit my access to the best healthcare.
12. I am on too many medications.
13. I am concerned about the side effects from my medications.
14. I am satisfied with the information my doctors share with me.
15. My doctors have limited management options to offer me.
16. My doctors spend adequate time with me.
17. My doctors are compassionate.
18. I have a say in the way my illness is managed.
19. My medications are working.
20. It is helpful to know the experiences of others with similar illness as mine.
21. It is unpleasant to use some of my medications.
22. It is physically difficult to handle some of my medications.
23. I vary my recommended management based on how I am feeling.
24. I put up with my medical problems before taking any action.
25. I push myself to follow the instructions of my doctors.
26. I get confused about my medications.
27. I make changes in the recommended management to suit my lifestyle.
28. I ensure I have enough medications so that I do not run out.
29. I have strict routines for using my regular medications.
30. I keep my medications close to where I need to use them.

Appendix G: Demographic Questionnaire

Completion of the demographic questionnaire is significant for determining the influence of variety of factors on the results of this study. All of these records will remain confidential. Please choose the appropriate answer.

1. What is your age?

◯   What is your age? 18-24

◯   25-34

◯   35-44

◯   45-54

◯   55-64

◯   65-74

◯   75 years or older

2. Are you Hispanic, Latino, or Spanish origin?

◯   Are you Hispanic, Latino, or Spanish origin? No, not of Hispanic ,Latino, or Spanish origin

◯   Yes, Mexican, Mexican Am., Chicano

◯   Yes, Puerto Rican

◯   Yes, Cuban

◯   Yes, another Hispanic, Latino, or Spanish origin, Type in origin:

3. What is your race?

◯   What is your race? Caucasian/White

○ Black/African American

○ Asian Indian

○ Japanese

○ Native Hawaiian

○ Chinese

○ Vietnamese

○ Samoan

○ Filipino

○ Korean

○ Guamanian or Chamorro

○ American Indian/Alaska Native

○ Other Asian

○ Other Pacific Islander

4. What is the highest level of education you have completed?

○ What is the highest level of education you have completed? Less than high school

○ High school graduate

○ Associate degree

○ Bachelor's degree

○ Master's degree

○ Ph.D, law or medical degree

5. Approximately what is your household income?

○   Approximately what is your household income? 10,000-19,999

○   20,000-29,999

○   30,000-39,999

○   40,000-49,999

○   50,000-59,999

○   60,000-69,999

○   70,000-79,999

○   80,000-89,999

○   90,000-99,999

○   100,000 or more

Appendix H: SGRQ

St. George's Respiratory Questionnaire
PART 1

Please describe how often your respiratory problems have affected you over the past 3 months.

Please check (✓) one box for each question:

	almost every day	several days a week	a few days a month	only with respiratory infections	not at all
1. Over the past 3 months, I have coughed:	☐	☐	☐	☐	☐
2. Over the past 3 months, I have brought up phlegm (sputum):	☐	☐	☐	☐	☐
3. Over the past 3 months, I have had shortness of breath:	☐	☐	☐	☐	☐
4. Over the past 3 months, I have had wheezing attacks:	☐	☐	☐	☐	☐

5. How many times during the past 3 months have you suffered from severe or very unpleasant respiratory attacks?

Please check (✓) one:

more than 3 times ☐
3 times ☐
2 times ☐
1 time ☐
none of the time ☐

6. How long did the worst respiratory attack last?
(Go to Question 7 if you did not have a severe attack)

Please check (✓) one:

a week or more ☐
3 or more days ☐
1 or 2 days ☐
less than a day ☐

7. Over the past 3 months, in a typical week, how many good days (with few respiratory problems) have you had?

Please check (✓) one:

No good days ☐
1 or 2 good days ☐
3 or 4 good days ☐
nearly every day was good ☐
every day was good ☐

8. If you wheeze, is it worse when you get up in the morning?

Please check (✓) one:

No ☐
Yes ☐

St. George's Respiratory Questionnaire
PART 2

Section 1

How would you describe your respiratory condition?

Please check (✓) *one*:

The most important problem I have	☐
Causes me quite a lot of problems	☐
Causes me a few problems	☐
Causes no problems	☐

If you have ever held a job:

Please check (✓) *one*:

My respiratory problems made me stop working altogether	☐
My respiratory problems interfere with my job or made me change my job	☐
My respiratory problems do not affect my job	☐

Section 2

These are questions about what activities usually make you feel short of breath these days.

For each statement please check
(✓) *the box* that applies
to you *these days*:

	True	False
Sitting or lying still	☐	☐
Washing or dressing yourself	☐	☐
Walking around the house	☐	☐
Walking outside on level ground	☐	☐
Walking up a flight of stairs	☐	☐
Walking up hills	☐	☐
Playing sports or other physical activities	☐	☐

St. George's Respiratory Questionnaire
PART 2

Section 3

These are more questions about your cough and shortness of breath <u>these days</u>.

For each statement please check
(✓) *the box* that applies
to you *these days*:

	True	False
Coughing hurts	☐	☐
Coughing makes me tired	☐	☐
I am short of breath when I talk	☐	☐
I am short of breath when I bend over	☐	☐
My coughing or breathing disturbs my sleep	☐	☐
I get exhausted easily	☐	☐

Section 4

These are questions about other effects that your respiratory problems may have on you <u>these days</u>.

For each statement, please
check (✓) *the box* that
applies to you *these days*:

	True	False
My cough or breathing is embarrassing in public	☐	☐
My respiratory problems are a nuisance to my family, friends or neighbors	☐	☐
I get afraid or panic when I cannot catch my breath	☐	☐
I feel that I am not in control of my respiratory problems	☐	☐
I do not expect my respiratory problems to get any better	☐	☐
I have become frail or an invalid because of my respiratory problems	☐	☐
Exercise is not safe for me	☐	☐
Everything seems too much of an effort	☐	☐

Section 5

These are questions about your respiratory treatment. If you are not receiving treatment go to section 6.

For each statement, please
check (✓) *the box* that applies
to you *these days*:

	True	False
My treatment does not help me very much	☐	☐
I get embarrassed using my medication in public	☐	☐
I have unpleasant side effects from my medication	☐	☐
My treatment interferes with my life a lot	☐	☐

USA / US English version   4

continued…

St. George's Respiratory Questionnaire
PART 2

Section 6

These are questions about how your activities might be affected by your respiratory problems.

For each statement, please check (✓)
the box that applies to you
because of your respiratory problems:

	True	False
I take a long time to get washed or dressed	☐	☐
I cannot take a bath or shower, or I take a long time to do it	☐	☐
I walk slower than other people my age, or I stop to rest	☐	☐
Jobs such as household chores take a long time, or I have to stop to rest	☐	☐
If I walk up one flight of stairs, I have to go slowly or stop	☐	☐
If I hurry or walk fast, I have to stop or slow down	☐	☐
My breathing makes it difficult to do things such as walk up hills, carry things up stairs, light gardening such as weeding, dance, bowl or play golf	☐	☐
My breathing makes it difficult to do things such as carry heavy loads, dig in the garden or shovel snow, jog or walk briskly (5 miles per hour), play tennis or swim	☐	☐
My breathing makes it difficult to do things such as very heavy manual work, ride a bike, run, swim fast, or play competitive sports	☐	☐

Section 7

We would like to know how your respiratory problems <u>usually</u> affect your daily life.

For each statement, please check (✓)
the box that applies to you *because of
your respiratory problems*:

	True	False
I cannot play sports or do other physical activities	☐	☐
I cannot go out for entertainment or recreation	☐	☐
I cannot go out of the house to do the shopping	☐	☐
I cannot do household chores	☐	☐
I cannot move far from my bed or chair	☐	☐

St. George's Respiratory Questionnaire

Here is a list of other activities that your respiratory problems may prevent you from doing. (You do not have to check these, they are just to remind you of ways your shortness of breath may affect you):

Going for walks or walking the dog

Doing activities or chores at home or in the garden

Sexual intercourse

Going to a place of worship, or a place of entertainment

Going out in bad weather or into smoky rooms

Visiting family or friends or playing with children

Please write in any other important activities that your respiratory problems may stop you from doing:

. . . .

. . . .

. . . .

. . . .

Now please check the box (one only) that you think best describes how your respiratory problems affect you:

It does not stop me from doing anything I would like to do ☐

It stops me from doing one or two things I would like to do ☐

It stops me from doing most of the things I would like to do ☐

It stops me from doing everything I would like to do ☐

Thank you for completing this questionnaire. Before you finish would you please make sure that you have answered all the questions.

Appendix I: Informed Consent

You are invited to take part in a research study of knowledge, attitude, lifestyle practices of Sporadic LAM patients and how it affects their quality of life. This study is being conducted by a researcher named Shahpar Vafamand, who is a doctoral student at Walden University. The researcher is inviting you to be in the study because you are a LAM patient. This form is part of a process called "informed consent" to allow you to understand this study before deciding whether to take part.

Background Information:

The purpose of this study is to measure the knowledge, attitude, and lifestyle practices of Sporadic LAM patients and how it affects their quality of life

Procedures:

If you agree to be in this study, you will be asked to participate in 5 questionnaires: demographic, knowledge, attitude, lifestyle practices, as well as quality of life. It should take up approximately 25-35 minutes of your time to complete the survey. Please do not include your name in any of the questionnaires you fill out since your identity will be kept confidential.

Voluntary Nature of the Study:

This study is voluntary. Everyone will respect your decision of whether or not you choose to be in the study. If you decide to join the study now, you can still change your mind later. You may stop at any time.

Risks and Benefits of Being in the Study:

Being in this study would not pose risk to your safety or wellbeing. Emotional upset may incure while answering the questions. The studies potential benefits may not directly benefit you, but the information learned about this study should provide more general benefit to the LAM community.

Payment:

There is no payment or compensations for participating in this study.

Privacy:

Any information you provide will be kept confidential. The researcher will not use your personal information for any purposes outside of this research project. Also, the researcher will not include your name or anything else that could identify you in the study reports. Data will be kept secure by the researcher only. Data will be kept for a period of at least 5 years, as required by the university.

Contacts and Questions:

You may ask any questions you have now or if you have questions later, you may contact the researcher via email at shahpar.vafamand@waldenu.edu or researcher's cellular phone at 703-579-7489. The researcher's advisor is Dr Amany Refaat who can be reached at amany.refaat@waldenu.edu . You can contact Walden representative with questions about your rights as participants at IRB@waldenu.edu or call them at 612-312-

1210. If you would like to receive a copy of the study results please indicate by checking

here_____

Walden University's approval number for this study is 06-13-14-0145430 and it expires

on June 12- 2015

Statement of Consent:

I have read the above information and I feel I understand the study well enough to make a

decision about my involvement. by clicking the link below.

_____ I understand that I am agreeing to the terms described above.

Appendix J: Permission to copy BCKQ

Subject:	RE: permission
From:	Roger White (roger.white4@virgin.net)
To:	shahparvafamand@yahoo.com;
Date:	Saturday, November 8, 2014 2:36 PM

Yes, Certainly you may. Glad to hear of your acheivement.

Roger White

From: Shahpar Vafamand [mailto:shahparvafamand@yahoo.com]
Sent: 08 November 2014 18:31
To: Roger White
Subject: permission

Hello Dr White,

My name is Shahpar Vafamand and I am about to graduate with my doctoral degree from Walden University. You were so gracious to allow me use the BCKQ for my study . I have to include a copy of the questionnaire in the appendix and wanted to get your permission before doing so. I would be extremely grateful. Thank you again for allowing me to use this valuable tool.

Shahpar

Appendix K: Permission to copy DLBQ

Subject: Re: permission

From: Lakerveld, Jeroen (je.lakerveld@vumc.nl)

To: shahparvafamand@yahoo.com;

Date: Wednesday, November 12, 2014 12:31 PM

Hi Shahpar,
No problem; you have my permission.
Best wishes,
Jeroen

Op 12 nov. 2014 om 18:27 heeft Shahpar Vafamand <shahparvafamand@yahoo.com> het volgende geschreven:

Hi Dr Lakerveld,

My name is Shahpar Vafamand and I am about to graduate with my doctoral degree from Walden University. You were so gracious to allow me use the DLBQ for my study . I have to include a copy of the questionnaire in the appendix and wanted to get your permission before doing so. I would be extremely grateful. Thank you again for allowing me to use this valuable tool.

Shahpar

Appendix M: permission to copy BBQ

Subject: FW: Permission

From: Johnson George (Johnson.George@monash.edu)

To: shahparvafamand@yahoo.com;

Date: Thursday, November 20, 2014 5:42 PM

Hi Shahpar

Thank you for your interest in the BBQ.

We are happy for you to use BBQ in your research with due acknowledgement to its developers and include a copy of the questionnaire in the appendix.

All the best.

Johnson

Dr Johnson George
Senior Lecturer
Faculty of Pharmacy and Pharmaceutical Sciences
Centre for Medicine Use and Safety

Monash University (Parkville Campus)
381 Royal Parade, Parkville
VIC 3052, Australia

Tel: +61-(0)3-9903 9178
Fax: +61-(0)3-9903 9629
E-mail: Johnson.George@monash.edu

www.pharm.monash.edu.au

From: Kay Stewart
Sent: Friday, 14 November 2014 1:09 PM
To: Johnson George
Subject: FW: Permission

Appendix N: Permission to copy SGRQ

St George's
University of London

Medicine, Biomedical Sciences, Health and Social Care Sciences

24 November 2014

Cranmer Terrace
London SW17 0RE

Switchboard
+44 (0)20 8672 9944

www.sgul.ac.uk

To Whom It May Concern:

This is to confirm that St George's, University of London (St George's Hospital Medical School) has given permission for Shahpar Vafamand, Walden University, Minneapolis, USA to use the St George's Respiratory Questionnaire (SGRQ) in a study entitled *"Knowledge, Attitude, Lifestyle Practices, and Quality of Life in Sporadic Lymphangioleiomyomatosis Patients".*

Professor Paul Jones, PhD FRCP
Professor of Respiratory Medicine

P.W. Jones, PhD FRCP
Professor of Respiratory Medicine

Curriculum Vitae

Shahpar Vafamand

Education:
PhD Public Health   Expected 2014
Walden University, Minneapolis, Minnesota
Dissertation Topic: Knowledge, attitude, lifestyle practices and quality of life in Sporadic
Lymphangioleiomyomatosis patients
Dissertation Chair: Dr. Amany Refaat

Masters of Science in Health and Physiology
George Mason University, Fairfax, Virginia   1994

Bachelor of Science in Environmental Health
Eastern Kentucky University, Richmond, Kentucky   1983

Work Experience:

Endocrinology Senior Sales Representative   2007-present
Abbott Pharmaceuticals, Lake Forest, Illinois
Responsible for the sales and marketing of Specialty Bio-Injection products to urology,
oncology, and gynecology market in Northern Virginia

Specialty Senior Sales Representative, Washington, DC   1998-2007
TAP Pharmaceuticals, Lake Forest, Illinois
Responsible for the sales and marketing of Specialty Bio-Injection product Lupron,
Urology, Oncology, and Lupron Gynecology in Washington, DC and Southern Maryland.

Accomplishments:

Continued market share increase in an extremely competitive market   2013-2011

Top 5% winning all Star Sales Achievement Award   2010
Winning the National Achievement Award for the number one spot in Urology market
share in the Nation

Top 10% in the Nation winning Excalibur Guild Award   2009

Top 10% in the Nation winning Excalibur Guild Award   2008

Top 10% in the Nation winning Excalibur Guild Award   2007

Top 10% in the Nation winning Excalibur Guild Award   2006

Exceed Urology goal in Q3 (107%)   2005
Led and outperformed the region and nation in Gynecology (Q2, Q3) (98%, 99% respectively)
Winner of t he 1st place in the discretionary contest incorporating gynecology volume growth with a details per day qualifier.
Chosen to represent and participate at the American Urological Associates yearly meeting
Chosen as the District trainer, successfully trained new hires.
I implemented district's Journal clubs and coaching sessions with district members.
I moderated successful learning via phone conferences with the District members.

Chosen as the Regional team leader for Kaiser Permanente   2004
Finished top 10% in the nation winning The Excalibur Guild Award (combined percent to goal, unit increase).
I achieved the winner for the nothing but net contest earning $5,000.00. I led the district in the new patient start on script assist earning $1,500.
 Leader in the district achieving goal in all four quarters of 2004 (106%, 103%, 114%, 110% respectively)
Increased Urology equivalent units sold in every quarter of 2004 by 8%.
Led the district in Gynecology finishing at 106% (Q1)
Winner of the second half Saiyushu Award (July-Dec combined percent to goal, unit increase)
Represented TAP at the American College of Obstetrics and Gynecology National meeting

Gained Senior Lupron Specialty Rep   2003
Gained a large competitor account all to Lupron (100 Zoladex patients)
Successfully Conducted 6 CME Credit programs with average attendees of 35 providers at each program.
Completed 8 CME Audio Conference (4 in Urology, 4 in Gynecology)
I was the winner of the Regional Award for the highest gynecology volume achieved in 2003 over 900 kits per quarter.
Winner of the spot awards for district input and district training.

Excalibur Guild Winner (Earning a trip to Rome)   2002
Winner of the Winner Circle Award

Outperformed Nation/Region/District in percent to goal, finishing over 109%
Winner of the second half Saiyushu Award
Winner of Home Stretch National Contest
Senior Representative Status
Winner of Drive to One Billion (BMW)
Winner of CHAS Award
Rated Exceeds with specific mention of solid closing skills, consistent teamwork, outstanding clinical skills, problem solving skills, product knowledge and decision making skills
As the District Trainer, successfully trained 3 new hires organized and developed the District Journal Club.
I was appointed as the member of the Region's Advisory Council.

Excalibur Guild Winner placing #4 in the nation (Earning a trip to Hawaii)   2001
Winner of 2nd half Saiyushu Award
I won the District's 4 month conversion contest (69.41%), higher than National/Regional average rate of 59% and 61% respectively.
I achieved the District's highest Gynecology Percent to goal and highest percent to goal increase over Quarter1.
Outperformed Nation/Region/District in Gynecology percent to goal attainment of 128% vs. 98%, 102% and 102% respectively

Outperformed Nation/Region/District in Urology percent to goal   2000
Attainment of 119% vs. 109%, 105% and 107% respectively

Excalibur Guild Award, placing 4th in the nation   1998-2000
1998 winner of 2nd and 3rd quarters District contests
1998 16% growth in Gynecology and 17% growth in Urology
1999 85% urology market share vs. 73% National average
2000 specialty sales representative

Chanel, Inc at Nordstrom, Arlington, VA   1989-1998
Product Specialist
Responsible for preparing and implementing promotional events as well as training seminars

Accomplishment:
Increased clientele base by 55% and achieved the number one sales/ promotion specialist within the region.

References available upon request