INTRODUCTION

Systemic Lupus Erythematosus (SLE or lupus) is a chronic inflammatory autoimmune disease of unknown etiology that commonly affects women of childbearing age. Similar to many other rheumatological diseases, it has a variable course and outcome and is subject to periods of exacerbation and remission. Frequently affecting the musculoskeletal system and skin, lupus can also cause inflammatory changes in the kidneys, lungs, heart and central nervous system. Lethargy and fatigue are described as the most disabling of Systemic Lupus Erythematosus symptoms***. (Balsamo & Santos, 2011).***

In SLE, the immune system that protects the body from viruses and bacteria malfunctions and generates antibodies that attack healthy tissue. As a result, inflammation can occur in the skin, muscles, joints, heart, lungs, kidneys, blood vessels and the nervous system. Even though lupus can affect men, women and children of any age. ***(EBertsias, Salmon & Boumpas, 2011).***

The cause of SLE is still unknown, but researchers believe it results from a combination of genetic and environmental factors. Some genes may predispose us to lupus, while others may protect us. Similarly, some environmental

factors may trigger the onset of lupus, while others may be protective. The onset of clinical SLE may therefore be attributed to one combination of specific genes and environmental factors in the absence of others. (***Perry, 2011.)***

Nursing role for patients with SLE is primarily depended on how to help those patients in adopting a healthy lifestyle. This includes healthy nutritional habits, exercising, keeping an ideal body weight, smoking cessation, healthy sleeping habits and a low-stress lifestyle. Additionally, medications will be needed to bring SLE symptoms under control, ideally bring about a remission and help the patient manage his/her disease. Early treatment can reduce permanent tissue damage and minimize the amount of time a person with SLE requires high doses of medications. ***(Stohl & Jacob, 2011).***

In Egypt it is reported that; about ٨٥cases diagnosed with SLE monthly are admitted to the rheumatology department with different signs and symptoms. ***(Information and Statistics Center of Ain Shams University Hospital, ٢٠١٢)***. SLE occurs nine times more often in women during their childbearing years (age 15 to 45) than in men. ***(David, 2011).***

Self management is essential to managing lupus. Self-management is what the client with SLE does to better manage their condition(s). Also it describes the strategies that

individuals use to manage the disease process itself, any emotional impacts of living with the condition, and the changes that occur to every day living as a result of the condition. This involves finding information, making decisions and taking action. (**Zhang, Hochberg, Perlmutter, Tan, Cohen & Medsger, *2011).***

Self-management does not mean that an individual must manage their own health without any medical or healthcare treatments or support. It does not necessarily mean less access to health care; sometimes it means more planned and directed care. It is a partnership between the patient and the healthcare professional with the aim of achieving desirable health outcomes. ***(Lorig, 2010).***

Lupus awareness is important for the patient to be educated about the symptoms of lupus in order to identify when flares are beginning. In the chronic phase of lupus, these symptoms may show up again and signal the start of another flare. The patient who notices signs of flare can bring them to the attention of the physician who will do a careful examination and order tests to check for other evidence. When caught at this stage, a small increase in the dosage of medication may be all that is necessary. ***(DeCastro, Morales & Wagner, 2011).***

SLE self-efficacy has been shown to be important in relation to human functioning in various areas, e.g., mental and physical health, human development, or coping with environmental hazards or burglary. Self efficacy is an important determinant of self -management behavior. Self- management involves a constant process of making behavioral choices and decisions. Self-efficacy expectations strongly influence these choices and decisions. Interventions to enhance self-management behavior and health functioning should be aimed at strengthening self-efficacy expectations. ***(Wang & Osmond, 2010).***

The fatigue of lupus can't be ignored. It's much more than just feeling tired. It can make getting out of bed every morning seem like climbing a mountain. Ordinary tasks, like cooking dinner or doing the laundry, can seem impossible. Fatigue is a symptom that others can't see, which means they may not understand how bad patient feels. Even worse, patient may feels like no matter how much get rest, the fatigue will never go away. With lupus, medical conditions such as chronic inflammation, anemia (a blood disorder), and certain medicines often make fatigue even worse. (***Long, 2010).***

With close follow-up and the right treatment, most people with lupus can expect to live a full and active life. However it can cause serious and even life-threatening

problems in some cases. Many people with lupus have ‘flares’, periods when their symptoms get worse. ‘Flares’ can happen with no obvious cause. There is no way of knowing their severity or how long they will last. They can occur more commonly during times of stress, or may be triggered by sun exposure, infections, and pregnancy. ***(Helve, 2010).***

Significance of the Study:

Systemic Lupus Erythematosus is a complex disease to diagnose, treat and manage. Patients should encourage taking control of their lupus, and managing it. It is important that patients are referred early for diagnosis and that they are also referred to members of the wider health professional team. Self-management and patient empowerment enable patients to make informed choices about their life ***(David, 2011).*** The systemic Lupus Erythematosus self-management guidelines had positive effects on the patients in reducing fatigue and depression and improving knowledge, coping skills and self-efficacy.

AIM OF THE STUDY

1. Assessing the needs of the patients with Systemic Lupus Erythmatosus.
2. Planning and implementing self management guidelines for patients with Systemic Lupus Erythmatosus.
3. Evaluating the effect of the guideline on patients with Systemic Lupus Erythmatosus self management.

Research Hypothesis:

The current study suggested four hypotheses:

1. The implementation of self management guideline will affect the needs of the patients with Systemic Lupus Erythmatosus

2. The implementation of self management guideline will improve the general self efficacy of the patients with Systemic Lupus Erythmatosus?

3. The implementation of self management guideline will decrease the fatigue severity among the patients with Systemic Lupus Erythmatosus?

4. The implementation of self management guideline will increase the patients' awareness' regarding the Systemic Lupus Erythmatosus?

REVIEW OF LITERATURE

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory disease that has protean manifestations and follows a relapsing and remitting course. SLE or lupus is a systemic autoimmune disease (or autoimmune connective tissue disease) that can affect any part of the body, but mainly involves the skin, joints, kidneys, blood cells, and nervous system. It is characterized by an autoantibody response to nuclear and cytoplasmic antigens. ***(Hodkinson, Musenge & Tikly, 2011).***

As occurs in other autoimmune diseases, the immune system attacks the body's cells and tissue, resulting in inflammation and tissue damage. It is a Type III hypersensitivity reaction caused by antibody-immune complex formation. ***(James, Watson, Gorgy, Tano & Mack, ٢٠٠٩).*** SLE most often harms the heart, joints, skin, lungs, blood vessels, liver, kidneys, and nervous system. The course of the disease is unpredictable, with periods of illness (called flares) alternating with remissions. ***(Syuto, Shimizu & Takeuchi, 2010).***

SLE can be fatal; there is currently no cure, although with recent medical advances, fatalities are becoming increasingly rare. However, lupus can he very successfully

treated with appropriate drugs and most persons with the disease can lead active, healthy lives. Survival for people with SLE in the United States, Canada, and Europe has risen to approximately 95% at five years, 90% at ١٠years, and 78% at ٢٠years. ***(Lupus foundation of America, 2011).***

Incidence and Prevalence of Lupus

Systemic Lupus Erythermatosus (SLE) is a disease of multifactorial etiology. The incidence and prevalence of SLE varies considerably across the countries. The burden of the disease is considerably elevated among non white racial groups. There is a trend towards higher incidence and prevalence of SLE in Europe and Australia compared to the USA. This variability may reflect true differences across populations. Also it's estimated that active SLE contributes to about a third of early deaths worldwide. ***(Seawell & Danoff, 2010).***

Lupus is most common in women of childbearing age, but it can occur in men and women of any age. Approximately 90% of patients with Systemic Lupus Erythematosus are women. The rate of SLE varies considerably between countries, ethnicity, gender, and changes over time. In the United States the prevalence of SLE is estimated to be about ٥٣per l٠O translating to about ١٥٩,٠٠٠out of ٣٠٠million people in the US being affected. In Europe the rate is about ٤٠ per ١٠٠,٠٠٠

people. The prevalence of lupus varies widely, ranging from ٤ cases to ٢٥٠ cases per ١٠٠,٠٠٠. ***(Lupus Foundation of America, ٢٠١١).***

Pathophysiolgy of SLE

SLE is an autoimmune disorder characterized by multisystem microvascular inflammation with the generation of auto-antibodies. Although the specific cause of SLE is unknown, multiple factors are associated with the development of the disease, including genetic, racial,. hormonal, and environmental factors. Many immune disturbances, both innate and acquired occurred in SLE. ***(Mendoza & Carrasco, 2011).***

In Systemic Lupus Erythematosus, many genetic-susceptibility factors, environmental triggers, antigen-antibody responses, B-cell and T-cell interactions, and immune clearance processes interact to generate and perpetuate autoimmunity. ***(Asanuma, Annette & Ayumi, 2009).***

Many clinical manifestations of SLE are mediated via circulating immune complexes in various tissues or the direct effects of antibodies to cell surface components. Immune complexes form in the microvasculature, leading to complement activation and inflammation. Moreover, antibody-antigen complexes deposit on the basement membranes of skin and kidneys. In active SLE, this process has been confirmed by

demonstration of complexes of nuclear antigens such as DNA, immunoglobulins, and complement proteins at these sites. ***(Paolo, 2009).***

Etiology of SLE

There is no one specific cause of Systemic Lupus Erythematosus. There are, however, a number of environmental triggers, genetic susceptibilities, and some drug reactions. ***(Rhodes & Vyse, 2008).***

1. Genetics triggers

The first mechanism may arise genetically. Research indicates SLE may have a genetic link. SLE does run in families, but no single causal gene has been identified. Instead, multiple genes appear to influence a person's chance of developing lupus when triggered by environmental factors. The most important genes are located in the HLA (human leukocyte antigen) region on chromosome ٦, where mutations may occur randomly or may be inherited. HLA class I, class II, and class III are associated with SLE, but only classes I and II contribute independently to increased risk of SLE. Also there are other genes which may contain risk variants for SLE such as, IRF ٥ (interferon regulatory factor), and STAT ٤(Signal Transducer and Activator of Transcription). (***Yang & Zhao, 2009).***

2. Environmental triggers

The second mechanism may be due to environmental factors. These factors may not only exacerbate existing SLE conditions, but also trigger the initial onset. Researchers have sought to find a connection between certain infectious agents (viruses and bacteria), but no pathogen can be consistently linked to the disease. ***(Graham, Barmak & Crow, 2010).***

3. Drug reactions

Drug-induced Lupus Erythematosus is a (generally) reversible condition that usually occurs in people being treated for a long-term illness. Drug-induced lupus mimics SLE. However, symptoms of drug-induced lupus generally disappear once the medication that triggered the episode is stopped. More than ٣٨medications can cause this condition, the most common of which are procainamide, isoniazid, hydralazine, quinidine, and phenytoin. ***(Geoffrey Horn, 2008).***

Types of SLE

Although lupus is used as a broad term, several kinds of lupus exist. There are four types of SLE as follows: ***The first and fatal type*** is; Systemic Lupus Erythematosus is the most common form of the disease. The symptoms of SLE may be mild or serious according the affected body organs. ***(Robert & Barmak, 2009).***

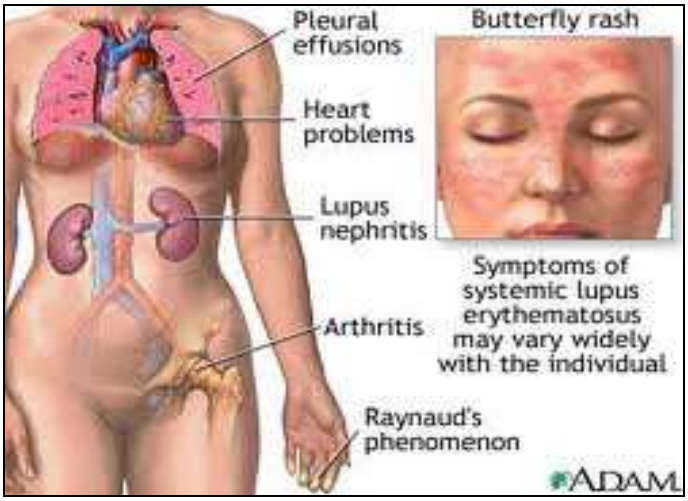
The second type is; Discoid Lupus Erythematosus (DLE) is a chronic skin condition of sores with inflammation and scarring favoring the face, ears, and scalp and at times on other body areas. These lesions develop as a red, inflamed patch with a scaling and crusty appearance. The center areas may appear lighter in color with a rim darker than the normal skin. When lesions occur in hairy areas such as the beard or scalp, permanent scarring and hair loss can occur. A small percentage of patients with discoid lupus can develop disease of the internal organs, which can make the person sick. ***(Geoffrey Horn, 2008).***

The third type is; Drug-induced Lupus Erythematosus (DILE); it is a variant of Lupus Erythematosus that resolves within days to months after withdrawal of the culprit drug in a patient with no underlying immune system dysfunction. DILE can arise months to years after exposure to drugs prescribed to treat various medical conditions (e.g. antihypertensives, antibiotics, anticonvulsants). The most common drugs that cause DILE are hydralazine, procainamide, quinidine, isoniazid, diltiazem, and minocycline. ***(Robert & Barmak, 2009).***

The last type is; Neonatal lupus is a rare form of lupus affecting newborn babies of women with SLE or certain other immune-system disorders. At birth, the babies have a skin rash, liver abnormalities, or low blood counts, which resolve entirely

over several months. However, babies with neonatal lupus may have a serious heart defect. Physicians can now identify most at-risk mothers, allowing for prompt treatment of the infant at or before birth. Neonatal lupus is very rare, and most infants of mothers with SLE are entirely healthy.***(Rhodes & Vyse, 2008).***

Signs and symptoms of SLE



Common symptoms of SLE. (Faizer, 2011)

Common initial and chronic complaints include fever, malaise, joint pains, myalgias, fatigue, and temporary loss of cognitive abilities. Because they are so often seen with other diseases, these signs and symptoms are not part of the diagnostic criteria for SLE. When occurring in conjunction with other signs and symptoms, however, they are considered suggestive. ***(Scheinfeld, DiCostanzo & Cohen, 2010).***

Dermatological manifestations. Many of patients with SLE sufferers of some de symptoms, with %٣٠to %٥٠ suffering from the classic malar rash (or butterfly rash) associated with the disease. Some may exhibit thick, red scaly patches on the skin (referred to as discoid lupus). Alopecia; mouth, nasal, urinary tract and vaginal ulcers, and lesions on the. skin are also possible manifestations. Tiny tears in delicate tissue around the eyes can occur after even minimal rubbing. ***(Gaipl, Kuhn & Sheriff, 2010).***

Musculoskeletal manifestations. The most commonly sought medical attention is for joint pain, with the small joints of the hand and wrist usually affected, although all joints are at risk. ***The Lupus Foundation of America (٢٠١٠)*** estimates more than 90% of those affected will experience joint and/or muscle pain at some time during the course of their illness. Unlike rheumatoid arthritis, lupus arthritis is less disabling and usually does not cause severe destruction of the joints ***(Hakkim, Fürnrohr & Amann, 2011).***

Fewer than ten percent of people with lupus arthritis will develop deformities of the hands and feet SLE patients are at particular risk of developing osteoarticular tuberculosis. A possible association between rheumatoid arthritis and SLE has been suggested, and SLE may be associated with an increased

risk of bone fractures in relatively young women. ***(Yasutomo, Horiuchi & Kagami, ٢٠٠9).***

Hematological manifestations. Anemia may develop in up to 50% of cases. Low platelet and white blood cell counts may be due to the disease or a side effect of pharmacological treatment. People with SLE may have an association with antiphospholipid antibody syndrome (a thrombotic disorder), where in autoantibodies to phospholipids are present in their serum. Abnormalities associated with antiphospholipid antibody syndrome include a paradoxical prolonged partial thromboplastin time (which usually occurs in hemorrhagic disorders) and a positive test for antiphospholipid antibodies; the combinations of such findings have earned the term "lupus anticoagulant-positive". Another autoantibody finding in SLE is the anticardiolipin antibody, which can cause a false positive test for syphilis. ***(Poole & Schneider, 2009).***

Cardiac manifestations. A person with SLE may have inflammation of various parts of the heart, such as pericarditis, myocarditis, and endocarditis. The endocarditis of SLE is characteristically non-infective (Libman-Sacks endocarditis), and involves either the mitral valve or the tricuspid valve. Atherosclerosis also tends to occur more often and advances more rapidly than in the general population. ***(Pan & Li WP, 2008).***