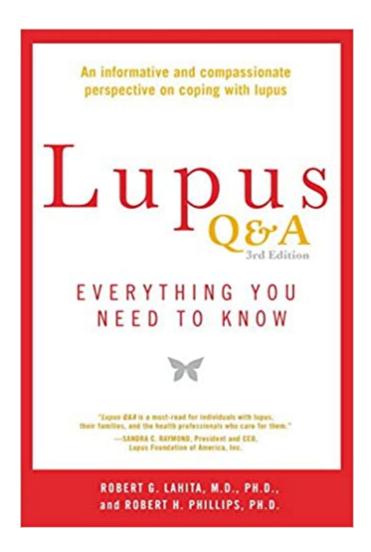


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Lupus Q&A Revised And Updated, 3rd Edition: Everything You Need To Know





Synopsis

A revised and updated edition of the bestselling resource for lupus patients, their families, and medical professionals \hat{A} \hat{A} A perennial bestseller, Lupus Q&A is the go-to guide for sufferers of a chronic autoimmune disease that affects more than 1.4 million people in the United States alone. Characterized by achy joints and skin rashes, lupus often mimics other diseases, making it tricky to diagnose and treat. In this completely revised and updated edition, Dr. Robert Lahita and Dr. Robert Phillips \hat{A} $\hat{\phi}$ \hat{a} $\hat{\phi}$ eleading experts on lupus \hat{A} $\hat{\phi}$ \hat{a} $\hat{\phi}$ ediscuss topics in a clear, concise, and easy-to-follow Q&A format. \hat{A} \hat{A} Lahita and Phillips review the newest drugs and explore beneficial complementary and alternative treatments, including new data on hormone use. Demystifying everything from diagnosis to the disease \hat{A} $\hat{\phi}$ \hat{a} $\hat{\phi}$ psychological impact, Lupus Q&A prepares readers to face the challenges ahead \hat{A} $\hat{\phi}$ \hat{a} \hat{a} and to restore their health and their lives.

Book Information

Paperback: 304 pages

Publisher: Avery; 3 Rev Upd edition (December 2, 2014)

Language: English

ISBN-10: 1583335455

ISBN-13: 978-1583335451

Product Dimensions: 6 x 0.8 x 9 inches

Shipping Weight: 12.6 ounces (View shipping rates and policies)

Average Customer Review: 4.8 out of 5 stars 6 customer reviews

Best Sellers Rank: #562,791 in Books (See Top 100 in Books) #16 inà Books > Health, Fitness & Dieting > Diseases & Physical Ailments > Lupus #35 inà Books > Health, Fitness & Dieting > Diseases & Physical Ailments > Rheumatic Diseases #49 inà Â Books > Health, Fitness & Dieting > Diseases & Physical Ailments > Reference

Customer Reviews

Robert G. Lahita, M.D., Ph.D., is the chairman of the Department of Medicine at Newark Beth Israel Medical Center. He has been internationally recognized for his clinical research in systemic lupus erythematosus and autoimmune disease, and is the recipient of numerous research grants from the Primus Foundation, the Lupus Foundation of America, and the National Institutes of Health. Dr. Lahita lives in Newark, New Jersey.Robert H. Phillips, Ph.D.,Ã Â a clinical psychologist, is the director of the Center for Coping on Long Island, NY. He is internationally recognized for his work helping people with chronic medical problems, is the author of more than 35 books, and has

presented at meetings all over the world. A former board member of the Lupus Foundation of America, he is on the national board of the American Autoimmune Related Diseases Association.

Original Foreword is my great pleasure to write a foreword to this book, dedicated to the patient with lupus and written by Dr. Robert Lahita, who has done so much for my family, in conjunction with Dr. Robert Phillips. Autoimmune disease, a topic that is very important to my family, can strike anyone at any age. One of the great mysteries of our time, this category of illness inflicts psychological and physical damage upon its sufferers. Systemic lupus erythematosus, in particular, has caused untold devastation and suffering for lupus patients and their families. It is also a disease that is harder to diagnose than it is to pronounce. This is why it is important to have a book like this oneA¢â ¬â •solid and comprehensive, yet simple and direct. It answers questions for patients, for their families, and perhaps even for their doctors. The authors have taken particular aim at the public interest in this disease that affects women most, but has an impact on people of every gender, of every age, and of every race, and in doing so have given some attention and interest to a disease that affects many but about which so little is known. I know, for this disease has touched, and will doubtless continue to touch, the members of my family. George joins me in wishing all patients and their families well and in hoping that Lupus Q & A: Everything You Need to Know will help many people to understand and cope with this dreaded disease. â⠬⠕BARBARA BUSHFormer First LadyHouston, TexasPrefaceThere is no more difficult disease to diagnose, understand, or treat than the disease called systemic lupus erythematosus. This may be because lupus is not one disease but many diseases grouped under one heading. It may also be because the disease can present itself to both physicians and patients in mysterious ways, throwing them off the track, leading them to think of other more common illnesses, and eluding standard diagnostic methods. Whatever the reason, lupus is complex and problematic. This book was designed to answer the many questions you may have about this disease and its impact on your life. Many of the questions are based on the countless numbers of patients who enter our clinics and offices daily with long lists of questions, the letters that arrive weekly in the mail seeking answers, the questions we hear at conventions or meetings, or the sad phone messages that ask for help because \tilde{A} ¢â ¬Å"my doctors don \tilde{A} ¢â ¬â,,¢t understand the disease. \tilde{A} ¢â ¬ \hat{A} • This is not a companion to any textbook. It is written strictly for patients and is based on their needs and questions. The goal was not to educate doctors with the material in this book (although many will find it helpful) but rather to address patients \hat{A} \hat{c} \hat{c} \hat{c} very real questions. We also realize that despite the dozens of questions answered in the book, certain areas may have been inadvertently overlooked. However, our goal

was to address questions that, according to our experience, are most on the minds of our patients. Ironically, the disease lupus seems to have gotten more complex, not less, over the years. New knowledge about the immune system and its workings has led to other autoimmune diseases being added to the roster of problems that need resolution. For example, because of difficulties in classification, a disease such as autoimmune phospholipid syndrome is often given the label 碉 ¬Å"lupus.Á¢â ¬Â• Some physicians label it lupus in order to give the disease a billing code acceptable for insurance reimbursement! Diseases such as autoimmune phospholipid syndrome have resulted in swelling numbers of lupus patients. The Lupus Foundation of America has estimated that some 2 to 2.5 million Americans believe that they have lupus, and that some 86 percent of Americans have heard of the disease. While these are staggering numbers, one must be cautioned to remember that they may reflect the fact that many illnesses that are not lupus are being called lupus. Hopefully, this book will help to clarify the reasons that the numbers of people with lupus are increasing. Last, a bit of history is necessary to allow readers to understand this disease and its past. Let $\tilde{A}\phi\hat{a} - \hat{a}_{\parallel}\phi$ s review the $\tilde{A}\phi\hat{a} - \hat{A}^{\dagger}$ timeline $\tilde{A}\phi\hat{a} - \hat{A} \cdot \phi$ of lupus.à à à Á Á c⠬¢Ã Lupus got its name because it was originally thought to represent the wounds that resulted from being attacked by a wolf. The trademark ââ ¬Å"butterfly rashâ⠬• was thought to be from the bite or scratches of a wolf.Ã Â Ã Â Ã Â Ã Â Á Å Å This butterfly rash on the malar part of the face (above the cheeks) was first mentioned in the thirteenth century. à à ¢â ¬Â¢Ã The actual term lupus erythematosus was first mentioned by a fellow named Cazenave in 1851. Ã Â Ã Â Ã Â Ã Â Ã Å There was much confusion regarding the diagnosis of lupus until well into the twentieth century. It was often confused with tuberculosis, disseminated gonorrhea, and many skin disorders. Ã Â Ã Â Ã Â Â Â Â Ô Ô Ô Ô Ô NI in the 1930s and 1940s did pathologists look at organs such as the kidneys and skin and realize that common changes in these organs had certain similarities in patients with lupus. Together these formed the typical aspects of lupus. \tilde{A} \hat{A} \tilde{A} \hat{A} $\hat{$ 1940s, and the classical description of antibody structure was not to take place for some decades. No one knew the mechanism through which lupus could so globally damage so many organs of the body. No one really understood this newly described illness, which counted rash, kidney failure, and sun sensitivity among its list of characteristics. Ã Â Â Â Â Â Â Â Â Â Â Â Â Î In the 1940s came the association with the false-positive test for syphilis, the discovery of the LE cell, and the idea that these phenomena might have something to do with \tilde{A} ¢â $\neg \tilde{A}$ "blood proteins. \tilde{A} ¢â $\neg \hat{A}$ • These proteins were later called antibodies, and it was suggested that they might be reacting with normal tissues. All of these discoveries were important in the understanding and diagnosis of

lupus.à à à Ácâ ¬Âcà Coincidentally, the discovery of cortisone in 1948 by Philip Hench provided the first and greatest therapy for lupus. \tilde{A} \hat{A} \tilde{A} \tilde{A} fluorescent antinuclear antibody assay, an important test in the diagnosis of lupus, and the discovery of autoantibodies such as Sm and RNP. These autoantibodies form the basis for our understanding of the disease process of lupus ¢â ¬â •how we diagnose it and a small bit about how the disease affects the body. In addition, the 1950s brought about important insight into the genetics of the disease. All of this added significantly to knowledge about lupus. Ã Â Ã Â Ã Â Á Á Since the 1950s, much research has focused on the following areas: molecular genetics (in order to learn more about the immune response), hormones and their importance, and, more recently, the development of several new drugs. Although lupus research has come far in the past twenty-five years, it will take an understanding of the cause of the diseaseâ⠬⠕currently unknownâ⠬⠕in order to develop a targeted cure. \tilde{A} \hat{A} \tilde{A} \tilde syndrome was described that in many cases is inexorably linked to lupus. It is troubling because it causes bleeding and clotting. It is usually a condition of $\tilde{A}\phi\hat{a} - \hat{A}$ sticky blood $\tilde{A}\phi\hat{a} - \hat{A}$ that can result in blood clots in the lungs or the brain. The addition of factors concerning the syndrome resulted in a revision of the criteria for the classification of lupus. Ã Â Ã Â Ã Â Â Â Â Â Â Â Â Î ln the first decade of the twenty-first century came a number of significant advances, which are detailed in this book. These include the use of newer biological agents for lupus such as belimumab and rituximab. Newer drugs previously used for transplant of organs came to use as well. Despite all of the history of this disease, we still have a long way to go in our understanding of lupus and related illnesses. However, we have made strides in the last thirty years that have revolutionized the way we examine many illnesses. Great discoveries are yet to happen. Let us hope that this book will continue to enlighten patients as we move ahead. A¢â ¬â •ROBERT LAHITA, M.D., PH.D.Newark Beth Israel Medical CenterRutgers, the New Jersey Medical SchoolNewark, New JerseyA¢â ¬â •ROBERT H. PHILLIPS, PH.D.Center for CopingLong Island, New York1Lupus: An OverviewLupus is a complex disease. So, of course, once patients are diagnosed with lupus, they have many guestions about this puzzling disorder. This chapter will introduce you to the basics of lupus. First, we will answer the question. ââ ¬Å"Just what is lupus?â⠬• We will then discuss the different types of lupus, the causes of the disease, and common myths surrounding the disorder. The Nature of LupusWHAT IS LUPUS?Lupus is an autoimmune disease. The immune system, which normally protects the body, turns against the body (auto) and attacks it. Lupus has no known cause and, as a result, no known cure. The disease can affect many different systems of the body, and there are many different ways

that it can affect people. WHY IS IT CALLED LUPUS? In the early twentieth century, most physicians thought lupus was a skin disease. The disease got its name because many patients looked as though they had been bitten or scratched by wolves. Lupus is Latin for \$\tilde{A}\psi a \tau \tilde{A}\psi wolf. \$\tilde{A}\psi a \tilde{A}\ IS LUPUS REFERRED TO AS A CHRONIC DISEASE? Lupus is considered a chronic disease because it is currently incurable. Once one experiences lupus symptoms, one has lupus forever. However, certain forms of lupusâ⠬⠕such as the drug-related formâ⠬⠕donââ ¬â,¢t fit into this incurable category because once the drugs that have $\tilde{A}\phi\hat{a} - \tilde{A}$ "caused $\tilde{A}\phi\hat{a} - \hat{A}$ • the lupus are withdrawn, the lupus symptoms go away. CAN LUPUS BE BOTH ACUTE AND CHRONIC? Yes. Lupus is a chronic disease but can have acute episodes. As a chronic disease, lupus often has a slow onset, and it is ongoing and incurable. The immune systemA¢â ¬â •unable to rid itself of what is perceived as a foreign substance \$\tilde{A}\psi a \tau \continues to react against the \$\tilde{A}\psi a \tau \tilde{A}\tilde{\text{foreign}} tissue.â⠬• This can be a significant problem, lasting for quite a whileâ⠬⠕possibly up to many months¢â ¬â •and can be accompanied by signs and symptoms of other chronic illnesses.But a patient can experience acute episodes, times when symptoms can worsen in immediate, abrupt, and occasionally severe ways. These acute manifestations, known as flares, can often occur this way, even in those who have had the disease for many years. It¢â ¬â,,¢s like a simmering pot of soup that all of a sudden bubbles over, or like a volcano that simmers and all of a sudden pops its lid but then goes back down and simmers for another twenty years. WHAT IS A CONNECTIVE TISSUE DISORDER? This is the former name for lupus and other related diseases. In the past, any disorder involving the muscles, tendons, and, in some cases, even bones, used to be referred to as a connective tissue disease. It was once believed that the inflammation of connective tissue occurred only from overuse. We now know that connective tissue can be inflamed for a variety of reasons, some of which have nothing to do with overuse.WHAT IS MIXED CONNECTIVE TISSUE DISEASE? Mixed connective tissue disease is referred to as an \tilde{A} ¢â ¬Å"overlap syndrome \tilde{A} ¢â ¬Â• because there seems to be an overlap of several different diseases or symptoms, suggesting more than one disease. There are no specific signs or symptoms of overlap disease. To help doctors make this diagnosis, mixed connective tissue disease was originally associated with the presence of an antibody or specific protein in the blood called ribonucleoprotein antibody (RNP). Some experts think that this overlap syndrome may be related more to a disease called scleroderma than to lupus. Many people with lupus have the anti-RNP antibody, but one must have very high titers, or strength, of the antibody for mixed connective disease to be present. So the best way to describe mixed connective tissue disease is to call it a hybrid illness. What Happens in Lupus WHAT EXACTLY HAPPENS IN LUPUS? It is difficult

to provide an accurate answer to this question. The key to understanding what happens in lupus is understanding the job of the immune system. The immune system is designed to protect the body from foreign invaders called antigens. The problem is that in lupus, the immune system cannot distinguish certain \tilde{A} ¢ \hat{a} ¬ \hat{A} "self \tilde{A} ¢ \hat{a} ¬ \hat{A} • tissues from foreign invaders and thus attacks inappropriate targets. So, for some reason, the immune system has a problem correctly identifying antigens and turns against the body that it is designed to protect. WHAT ARE THE MAIN COMPONENTS OF THE IMMUNE SYSTEM? The immune system is primarily made up of three categories of cells¢â ¬â •B lymphocytes (commonly called B cells), T lymphocytes (T cells), and phagocytes. These three groups of cells form the $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ soldiers $\tilde{A}\phi\hat{a}$ $\neg \hat{A} \cdot$ of the immune system, designed to protect the body from foreign invaders (antigens). WHAT EXACTLY IS AN ANTIGEN? Antigens are any substances, produced inside the body or coming from outside the body, that the immune system recognizes as being foreign¢â ¬â •such as germs, bacteria, viruses, or fungi. In other words, an antigen is any substance that can trigger an immune responseâ⠬⠕an effort made by the immune system to eliminate an unwanted foreign invader. In lupus, the immune system suddenly and for no apparent reason turns against the body $\tilde{A}\phi \hat{a} - \hat{a}_{,,\phi} \phi \hat{a}$ own cells and tissues, mistaking them for foreign invaders, and tries to destroy them as it would other foreign matter. So the body is now reacting to autoantigens (self-antigens). WHAT IS AN AUTOANTIGEN? An autoantigen is a substance that occurs naturally within the body \tilde{A} ¢ \hat{a} $\neg \hat{a}$ •in other words, it is not really foreign but is a $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ self $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ substance $\tilde{A}\phi\hat{a}$ $\neg \hat{a}$ but, for some reason, is identified by the immune system as foreign. The autoantigen then triggers an immune response, and the body fights it the way it would normally fight off foreign substances with antibodies. This is called an autoimmune response. WHAT ARE ANTIBODIES? Antibodies are proteins that are among the five major classes of immunoglobulin molecules, protein molecules that work to kill foreign substances. They originate in the immune system¢â ¬â,,¢s B cells in response to the presence of the foreign substances (antigens), for the primary purpose of destroying them. B cells mature to become plasma cells, or those cells that make antibodies. WHAT ARE AUTOANTIBODIES? As you now know, antibodies are molecules that normally defend the body against foreign substances. Autoantibodies are the names for antibodies that fight healthy tissues within the body. No two patients have autoantibodies that have exactly the same target, although in families, two members with the disease may have autoantibodies that look very similar.WHAT DO ANTIBODIES DO?Antibodies attach themselves to foreign substances until the combination of antibody and foreign substance (the immune complex) is engulfed by a scavenger cell, called a phagocyte. This cell usually digests the immune complex, destroying it. In some cases, after the immune complex is

engulfed, the antibody and foreign agent go directly to the spleen to be deposited. The spleen is the depository or $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ "graveyard $\tilde{A}\phi\hat{a}$ $\neg \hat{A} \cdot$ of old immune complexes. There are also cells in the spleen that are capable of engulfing and removing substances from circulation themselves.WHAT IS AN ANTIGEN-ANTIBODY REACTION? The antigen-antibody reaction is the process in the immune system that creates immune complexes. The white blood cells (consisting of B cells that become cells that make antibodies) respond to an antigen. The antibody then binds with the antigen. An antigen is usually a foreign bacterium or virus in a healthy person. In a lupus patient, an antigen might be anything, such as a clotting factor or a white cell.WHAT EXACTLY IS AN IMMUNE COMPLEX? Very simply, when the immune system reacts to an antigen and antibodies attack it to destroy it, the attachment of antibody to antigen is called an immune complex.WHAT ARE LYMPHOCYTES?Lymphocytes are the part of the white blood cell family that are produced in lymphoid tissue. They are the main cells of the immune system. There are different categories of lymphocytes, such as killer cells (described shortly) and T and B cells. All of these cells have a purpose, such as attacking and swallowing foreign materials, eliciting antibodies, or killing virus particles. These cells have amazing capabilities, and new functions are being discovered every year. Since lupus is a disease of the immune system, lymphocytes play a major role¢â ¬â •both in the manifestation of the disease and in the actual cause of illness. They have many roles: They make antibodies, they recognize foreign invaders, and they secrete chemicals that produce fever and other symptoms of the disease. WHAT IS NEW WITH THE IMMUNE SYSTEM? In order to understand the newer developments in lupus we have to understand that the immune system has been divided into two parts, the adaptive and the innate immune system. The adaptive immune system is common to humans and involves the interaction of cells such as T and B cells and the origins of specific antibodies directed toward foreign invaders. There is also something called the innate immune system, which is quite primitive and is present in insects and animals that have no backbone. These two aspects of immunity are critical to normal function, and they play a major role in the onset and promulgation of lupus. Both are under intense study. WHAT ARE T CELLS? The term T cells is shorthand for thymus-derived lymphocytes. There are several varieties of T cells. They can be $\tilde{A}\phi\hat{a} - \tilde{A}$ "helper $\tilde{A}\phi\hat{a} - \hat{A}$ cells (called helper T cells) that help immune function by alerting the B cells to begin producing antibodies to fight off the antigens, \tilde{A} ¢â $\neg \hat{A}$ "suppressor \tilde{A} ¢â $\neg \hat{A}$ • cells that suppress immune function, or killer T cells (also called cytotoxic T cells) that recognize, attack, and destroy antigens. More recently, T cells have been shown to regulate the functions of a variety of cells throughout the body, some not even considered

immune. They are called T regulatory cells, and they are being targeted to control lupus progression

and some of the symptoms. Most T cells have certain markers on their surfaces, called cluster determinants or CD markers. They are usually given a number. There are about 350 CD markers. Many researchers have CD marker maps on their office walls because the markers are impossible to remember. These markers are important because they help us find subsets of lymphocytes using special machines that sort the cells. The subsets of lymphocytes are very important to know in a particular patient. The identification of subsets is important in diagnosis and treatment of a variety of diseases, not just lupus. CD markers were discovered for the first time with specific monoclonal antibodies (antibodies that are $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ "tailor made $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ • in a laboratory in a mouse or in a cell culture). Monoclonal antibodies are not present in people with lupus. The monoclonal antibody is a way of learning more about these T cells in the laboratory. Essentially, the antibody was produced from an engineered fusion of two kinds of cells \tilde{A} ¢ \hat{a} $\neg \hat{a}$ •a cancer cell and a normal immune cell. These antibodies are highly specific and make great chemicals for study in the laboratory. Their \tilde{A} ¢â ¬Å"discoverers \tilde{A} ¢â ¬Â• were even awarded a Nobel Prize.We will speak about the use of these monoclonal antibodies in the treatment of disease later in the book.WHAT ARE B CELLS?B cells (which received their name because they were discovered in the bursa part of the chicken gut) are specialized lymphocytes that are responsible for a variety of functions. Their most important role, however, is their maturation into plasma cells and their eventual production of antibodies. B cells have a major role in the cause and manifestations of the disease lupus.DO B CELLS COME FROM THE BURSA IN HUMANS?No, but they are believed to come from the bone marrow, which is comparable to the bursa in humans. HOW DOES THE IMMUNE SYSTEM PRODUCE ANTIBODIES? The immune system is programmed shortly after birth, or perhaps even before, to recognize certain antigens as self-antigens. If a person is later exposed to an antigen that is not recognized as self, his or her immune system will reject it as foreign. This process of recognition goes on throughout life. When a person becomes infected with any organism, an immune response is made to that organism. After the initial reaction, there continues to be immunological memory for this foreign substance. One goal of the immune system is the production of killer cells that are designed to counter any invasion of foreign viral materials. Another goal is the manufacture of antibodies. Antibodies engulf foreign substances and remove them from circulation via the formation of immune complexes. The immune system includes T cells and B cells. B cells are responsible for making antibody. They can be stimulated to make antibody either by the foreign substance directly or by T cells. When B cells make antibody in response to stimulation by foreign matter, it is called a T-independent response. However, T cells, also called helper or suppressor cells, can come along and stimulate the B cells to make more antibody. These are called T-dependent responses. WHAT

DO T AND B CELLS DO?T cells recognize foreign invaders and respond by either killing them outright (\tilde{A} ¢â $\neg \hat{A}$ "killer \tilde{A} ¢â $\neg \hat{A}$ • T cells) or by presenting them to B cells so that a specific antibody can be made. DESCRIBE THE RELATIONSHIP BETWEEN T LYMPHOCYTES AND B LYMPHOCYTES. This relationship is one of the most important ones of the immune system. T cells remember previously identified antigens and usually activate themselves and B cells. Antigens are usually recognized by what are called antigen-presenting cells (APCs) that are strategically placed in such parts of the body as the lungs, liver, and prostate. APCs present the information, derived from a complex process of recognition, to T cells. The T cell never forgets an antigen. In a more complicated series of steps, antigens can also be directly presented to B cells. When a T cell is activated, it activates B cells to respond to the antigen by specializing, or turning their efforts toward that foreign substance. This process, called differentiation, allows the B cells to make antibodies that target the antigens. The amazing nature of this response has dazzled scientists for years and is still not fully understood. This half of the immune system, involving B and T cells, is called the adaptive immune system because it involves memory and the making of antibodies in response to memory of antigens. ARE THERE OTHER NEW AND IMPORTANT CELLS IN THE IMMUNE SYSTEM? Some cells that have major importance to the immune system are called dendritic (Greek for $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ "tree $\tilde{A}\phi\hat{a}$ $\neg \hat{A}$ oclls, because they look like they have branches as on trees. These cells are part of the innate immune system; they produce chemicals in response to a \tilde{A} ¢â $\neg \tilde{A}$ "happening, \tilde{A} ¢â $\neg \tilde{A}$ • as in the breach of a security system. Dendritic cells are present in all areas of the body such as the skin, nose, and lungs. Remember that the immune system is there to protect you from foreign invaders. Well, it is more sophisticated than previously thought, and the innate immune system is an important part of the alarm that tells your body to prepare for war on foreign invaders. Macrophages (APCs, described earlier) are part of this system.CAN THESE CELLS BE PART OF THE LUPUS DISEASE? These cells probably have more importance to lupus than previously thought. They may be the cells that $\tilde{A}\phi\hat{a} - \tilde{A}$ sound the alarm $\tilde{A}\phi\hat{a} - \hat{A} \cdot to$ warn T and B cells and activate interactions to protect the body.WHAT ARE PHAGOCYTES?Phagocytes (also called accessory cells because they assist in the immune response) are white blood cells that can destroy certain particles or debris. The name phagocyte comes from the Greek phagos, meaning \tilde{A} ¢â ¬Å"to eat. \tilde{A} ¢â ¬Â•WHAT ARE MACROPHAGES?Macrophages are large, mature phagocytes that are part of the innate immune system mentioned earlier. They have a number of important functions. They can, without any additional signal from the immune system, ingest and destroy foreign invaders, diseased cells, or cellular debris. They send signals to lymphocytes to alert them that antigens are present. They can also produce different cytokines (messengers to the immune

system that facilitate the immune response). WHAT IS COMPLEMENT? Complements are special proteins in the blood that help the antibodies in their effort to rid the body of the antibody-antigen combination (the immune complex). They are so named because they complement, or enhance, the function of the immune system. When an antigen is identified by the immune system as requiring destruction and an antibody attaches to it, this triggers a chain reaction called a complement cascade. In this chain reaction, a series of complement components that are normally inactive in the blood become activated and help the antibody eliminate the antigen. A category of markers for lupus are called complement components. There are nine components of complement, which as a whole serve as amplifiers of the immune reaction. In other words, complement makes the immune reaction more efficient. Deficiencies in any of the nine complement components result in a derailment of efficient immune system functioning. (We discuss complement in more detail in Chapter 2.) For some reason, when a person is deficient in some of the early components of complement, a lupuslike condition can be acquired. Such components include C2, C4, and C1Q.DOES MY DOCTOR MEASURE CERTAIN COMPLEMENT COMPONENTS TO LOOK AT LUPUS ACTIVITY? Remember, complement is a kind of amplifier of the immune reaction and as such must be examined when a patient sees the doctor for an evaluation. Typically the doctor will look at some parts of the cascade. The measurement of C3, C4, and possibly CH50 will help your doctor understand whether immune complexes are forming. The complement cascade has nine parts, and each is important.WHAT ARE LEUKOCYTES?Leukocytes are white blood cells (including such cells as phagocytes, lymphocytes, and monocytes). This is a broad term for many of the cells already discussed. When they receive the signal of invasion, they secrete chemicals in an attempt to kill the antigens that cause the joint pain and swelling when a lupus patient goes into a flare. They can also be involved in the process of destroying foreign invaders. Leukocytes can be involved in causing many problems in lupus, such as pulmonary hypertension, rashes on the face, or the breakdown of lung tissue. They can also be stimulated by a variety of agents or complexes (such as immune complexes). WHY IS LUPUS AN INFLAMMATORY DISEASE? Inflammation is a reaction of tissues to infection, injury, or invasion. It is usually characterized by swelling, redness, pain, heat, and reduced function. In lupus, inflammation is usually a result of an immune system reaction. The immune system believes that one of the otherwise healthy tissues in the body is the foreign invader and reacts accordingly, triggering the immune reaction that results in inflammation. HOW DOES INFLAMMATION OCCUR?Inflammation is the result of a series of very well-engineered chemical reactions, involving many molecules called mediators. An immune reaction occurs when an antigen is recognized. The white blood cells, specifically the lymphocytes, generally trigger a signal that is

sent to cells that contain the mediators. This signal triggers the release of the mediators. The powerful mediators then cause all of the pain, redness, and shifts in fluid and some of the swelling that occurs around swollen joints and in the skin that we typically recognize as inflammation. These symptoms are all indicators that the immune system is attacking and ingesting the antigens. For example, the rash on the cheeks typical in lupus patients, called the malar rash, is an effect of these inflammatory mediators.CAN YOU EXPLAIN THE INFLAMMATION PROCESS IN A LITTLE MORE DETAIL?Inflammation is caused by any insult or injury to a tissue or to a cell. The injury could be caused by pressure, trauma, heat, cold, or an organism¢â ¬â •whatever. It sets off an alarm, which activates a variety of cells. The cells come from the immune system and the bone marrow. The immune system cells are the lymphocytes and the macrophages. They compose the two large halves of the immune system. The macrophage (formally called a part of the innate immune system) is an important cell in this process. As APCs, the macrophages \hat{A} ¢â $\neg \hat{A}$ "register \hat{A} ¢â $\neg \hat{A}$ • the injury and determine whether there is any foreign material at the site of the injury. When the macrophages determine that the material is foreign, they transmit a chemical code to the lymphocytes saying that this is a foreign invader and give detailed information about the invader, such as how many amino acids it has, what it looks like, and whether it¢â ¬â,¢s a fat, a sugar, a protein, and so on. It gives the immune system the memory to recognize this invader when it enters the body again. Other cells that are important and related to macrophages are the various dendritic cells that assist in mounting an alarm through the secretion of chemicals that alert the other parts of the immune system. The bone marrow cells and those in the lymph nodes are leukocytes. They are part of the adaptive immune system and comprise phagocytes (the eater cells), T cells, and B cells. The phagocytes come along and scoop up the foreign materials or the debris caused by the injury. These are the inflammatory cells. They come to the scene and release chemicals that cause swelling, inflammation, and pain. The area is walled off. ItA¢â ¬â,¢s flooded with fluids. And all of that inflammation, pain, swelling, and fluid goes away when everything is cleaned up. The T and B cells are involved in the recognition of the invaders and the formation of antibody. WHY IS THERE REDNESS AND PAIN WITH INFLAMMATION? It is likely to be nature¢â ¬â, ¢s way of protecting the area. It prevents you from further damaging the area that is already inflamed and painful. Your tendency is to not feel, rub, touch, or do anything to the area that is inflamed. Inflammation walls the area off from normal cells. An analogy would be putting yellow tape around a crime scene. It $\hat{A}\phi\hat{a} - \hat{a}_{,\phi}\phi$ s to surround the area, to block off blood vessels. This blocking of the blood vessels causes the area to get red and painful. As far as the body is concerned, the pain is a minor aspect. It plays no essential role in clearing the damage. The pain is

strictly for the organism \tilde{A} ¢ \hat{a} $\neg \hat{a}$,¢s brain to prevent the organism from playing around, touching. rubbing, licking, or doing whatever else it might do to the area that might impede the healing process.WHAT ARE SOME OF THE CHEMICALS INVOLVED IN INFLAMMATION? Prostaglandins and leukotrienes produce pain and swelling in various areas of the body and account for the migration of cells to an inflamed area. Drugs such as aspirin that are used to control the inflammatory response directly affect these chemicals in order to decrease inflammation. The prostaglandins are a family of biologically active fats that are formed as a result of the action of an enzyme commonly known as a cyclooxygenase, or COX (see the discussion of drug therapy here). The most important action of the prostaglandins is the regulation of the inflammatory response. The leukotrienes are another group of chemicals that do a number of things during the inflammatory response. Among the functions of the leukotrienes is their ability to bring white cells to an inflamed area and to increase the permeability of blood vessels, which accounts for swelling and redness. However, the guide function is part of a group of chemicals now known as chemokines.WHAT IS PUS?Pus is nothing more than dead white cells \tilde{A} ¢ \hat{a} $\neg \hat{a}$ •cells that have come to the area and participated in the inflammatory/destruction process.WHERE DOES INFLAMMATION OCCUR?Inflammation can occur anywhere in the body. Most commonly, however, it occurs in joint spaces and other areas where there are white cells and immune complexes. Almost any organ can become inflamed from the heart to the brain. DOES INFLAMMATION HAVE A NEW ROLE IN LUPUS?Yes, scientists are now looking at something called the inflammasome, which is responsible for activation of the inflammatory response and the perpetuation of lupus, the patterns and numbers of flares that patients experience, and the fatigue and weakness experienced by most people.

Noce

Very helpful for care-giver as well.

this book has a lot of merit and information, however, for me, the q & a format gets tedious.

Very comforting to read

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Very good product

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