Lupus and Epidemiology: A Backgrounder

What is Lupus?

Lupus is an unpredictable and misunderstood autoimmune disease that ravages different parts of the body. It is difficult to diagnose, hard to live with, and a challenge to treat. Lupus is mostly hidden from view and undefined; a range of symptoms, strikes without warning, and has no known cause and no known cure. Its health effects can range from a skin rash to a heart attack. Symptoms of lupus come and go, change over time, and often imitate other illnesses, making lupus difficult to diagnose. Common symptoms include joint pain, skin rashes, overwhelming fatigue, and fevers that last for days or weeks. The exact cause of lupus is unknown.

Are there Different Forms of Lupus?

When people refer to ‘lupus’ they often mean systemic lupus erythematosus (SLE), which is the most common form of lupus. Systemic lupus can affect any part of the body, including the skin, joints, connective tissue, heart, lungs, kidneys, brain, blood vessels and blood. Health effects can range from mild to life-threatening and may include heart attacks, strokes, seizures, and miscarriages.

Cutaneous lupus erythematosus (CLE) is limited to the skin. Although cutaneous lupus can cause many types of rashes and lesions (sores), the most common—called discoid rash—is raised, scaly and red, but not itchy. Areas of rash appear like disks, or circles. In some cases, cutaneous lupus can evolve into systemic lupus.

Drug-induced lupus (DIL) is a lupus-like disease caused by certain prescription drugs. The symptoms of drug-induced lupus are similar to those of systemic lupus, but it rarely affects major organs.

Neonatal lupus is a rare condition associated with anti-SSA/Ro and/or anti-SSB/La antibodies from the mother that affect the fetus. At birth, the baby may have a skin rash, liver problems, or low blood cell counts, but these symptoms typically disappear completely after six months with no lasting effects.

Why is Lupus so Difficult to Diagnose?

There are many challenges to reaching a lupus diagnosis. Lupus symptoms mimic many other illnesses. The symptoms also can be unclear, can come and go, and can change over time. There is no single test that can determine whether a person definitely has lupus. Test results that suggest lupus can be due to other illnesses and can be seen in healthy people. A test result may be positive one time and negative another time. Different laboratories may produce different test results.
**Why Conduct a National Epidemiology Study on Lupus?**

In order to better understand the scope of the disease, scientists want to know **incidence**: how many people develop lupus each year; and **prevalence**: how many people in total are living with the disease. They also need to know if certain groups of people develop lupus more frequently than others. Investigators also want to determine whether there are patterns that will help predict which groups are at greater risk for the disease and to determine the **burden of disease** on specific populations. Having good epidemiological data on lupus also helps to stimulate public and private investment in lupus research by providing industry leaders and policymakers reliable information on the needs of people affected by lupus.

**What is the National Lupus Patient Registry?**

This title describes a U.S. Centers for Disease Control and Prevention project that funds five population-based lupus registries to measure the true magnitude and impact of lupus and better define the incidence and prevalence of lupus among all populations at risk for developing lupus. The sites are Georgia, Michigan, New York, California and the Indian Health Service. Funding for the registries was provided by the U.S. Congress in response to advocacy efforts by the Lupus Foundation of America.

**Have There Been Other Epidemiological Studies in Lupus?**

There have been a number of epidemiological studies in lupus over the past several decades. However, previous studies on lupus had limitations. Many were conducted at specific medical centers, which involved a small group of patients from areas whose populations lacked diversity or did not have adequate representation by all racial and ethnic groups at risk for lupus. These studies may have used outdated criteria, did not utilize common methodology, and did not include all forms of lupus. There is a need to obtain more complete data on the number of people affected by all forms of lupus using a common methodology.

**What is the Burden of the Disease for People Living with Lupus?**

The physical, emotional, and financial burden of living with lupus is staggering. A recent Roper survey revealed that 68 percent of people with lupus say the disease affects every relationship and their ability to perform daily responsibilities. Many people with lupus are not able to work or need to reduce the hours they do work. In fact, research has shown that only 31 percent of Americans with lupus report being employed full-time. According to a study published in 2009, the total mean annual direct and indirect costs to employers per patient with lupus was $21,215 and the total average mean direct and indirect costs to employers per patient with lupus-related kidney disease (lupus nephritis) was $64,195. (J Occup Environ Med. 2009;51:66–79).

**How Do These Two Reports Differ From Other Lupus Epidemiology Studies?**

Under the direction of the U.S. Centers for Disease Control and Prevention (CDC), in partnership with the Georgia and Michigan state health departments, investigators from Emory University and the University of Michigan led these landmark epidemiology studies using common methods and target populations.
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This project developed population-based registries to better define the incidence and prevalence of lupus (both systemic lupus erythematosus and discoid lupus) and better characterize individuals with these conditions.

Project Objectives

- Determine how many cases of lupus exist.
- Determine how many new cases occur each year.

Existing registries in Georgia and Michigan are focused on developing estimates for white and black populations. Newer registries exist in California and New York City will provide similar estimates for Hispanics and Asians, and in the Indian Health Service for American Indians/Alaska Natives. This data will become available in 2015.

What Did We Learn?

There are substantial racial disparities in the incidence of lupus and the new data finds higher rates of prevalence than found in most previous studies.

- Black women living with lupus were diagnosed with lupus at a younger age compared to whites, and had a higher proportion of renal disease and end-stage renal disease.
- Black women had very high rates of lupus, with an incidence rate nearly three times higher than that for white women, with significantly high rates in the 30-39 age group.

Note: At this time, it is not possible to extrapolate a national incidence and prevalence of lupus using only data from these two sites. Additional sites will report their data as it becomes available in the coming years.

What Were the Specific Findings for Each State?

**Incidence Rates**

In Georgia, the overall age-adjusted incidence rate was 5.6 per 100,000 person years. The rate for women was more than five times greater than for men (9.2 vs. 1.8). Black women had an incidence rates nearly three times higher than that for white women (13.4 vs. 4.7). The overall age-adjusted incidence rate in Michigan was 5.5. Also, the rate for women was higher than for men (9.3 vs. 1.5) and the rate for black women was more than twice that for white women (12.8 vs. 6.3).

**Prevalence Rates**

The overall age-adjusted prevalence in Georgia was 73.0 per 100,000 and 72.8 in Michigan. The prevalence for women in Georgia and Michigan was 127.6 and 128.7 respectively. Blacks were three times more likely to have lupus than whites in both Georgia (118.5 vs. 32.7) and Michigan (111.6 vs. 47.) The highest prevalence in both Georgia and Michigan was among black females (196.2 and 186.3 respectively).
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**Racial Disparities**

The Michigan investigators found substantial racial disparities in the burden of lupus, with blacks experiencing earlier diagnosis, more than two times the incidence and prevalence, and an increased number of people having their kidneys affected and progression to end stage renal disease (ESRD). The same disparity was seen by investigators in Georgia who found a striking difference in the proportion with ESRD, with a seven-fold greater involvement among blacks. They also noted a similar higher incidence among young black women, especially in the 30-39 age group.

**What’s Next?**

The investigators plan to use their lupus patient registries to conduct ongoing studies to document the progression of the disease and the economic burden of lupus over time, which, according to data analysis already available, is substantial.

Through improved management of the disease, people with lupus now have increased survival rates, but many will face a lifetime of serious health problems that will require expensive medical care, citing the need for increased investment in lupus research and development of new and more targeted therapies to bring the disease under control and improve quality of life.

Preliminary data from the Indian Health Service study will be presented during the American College of Rheumatology Annual Scientific Meeting, October 25-30, 2013 in San Diego, CA. Data from the New York and California sites are expected to be published sometime during 2015.