Study of Systemic Lupus Erythematosus

Systemic lupus erythematosus, or SLE, is to be the subject of a special controlled study at five of the Arthritis Clinical Study Centers supported by the National Foundation-March of Dimes.

The study is designed to obtain information about the course of the disease and to measure the value of treatment procedures, especially of high corticosteroid therapy, in saving and prolonging the lives of SLE patients with kidney involvement.

SLE is considered to be a "collagen disease," along with polyarteritis nodosa, scleroderma, dermatomyositis, rheumatic fever, and rheumatoid arthritis. The disease affects the connective tissues and almost always causes arthritis in the joints.

Dr. William S. Clark, director of medical care of the National Foundation, has stated that "until recently, lupus (or SLE) was generally thought to be invariably fatal. . . . This simply isn't so. In some forms it is never fatal. And even when kidney disease is present, doctors are beginning to find that they can keep many of their patients alive and even in reasonably good health for many years. . . . The big problem is that the disease follows such an unpredictable course, with ups and downs in severity and variations in reactions to treatment by different patients."

Although the true incidence of SLE is unknown, the number of cases being diagnosed as a result of improved procedures indicates that the disease is "relatively common." Dr. Clark believes that if mild and unrecognized cases are included, the annual incidence of SLE in the United States may be "upwards of 5,000."

Most victims of SLE are women, and, although the disease may have its onset at any age, it most often begins during the third and fourth decades of life. It may come and go, alternating acute periods with periods of remission, and may affect additional systems when it recurs, or it may be acute and result in death within a few weeks.

The cause of SLE is unknown, but autoimmune reactions are suspected because of certain blood protein abnormalities associated with the disease. The possibility that SLE may be the result of toxic or allergic reactions to drugs is being studied intensively. For example, when hydralazine is used to treat hypertension it sometimes produces many symptoms of SLE, and when the drug is stopped the syndrome disappears. The possibility that infection with bacteria or virus may be the cause of SLE is also being studied. Endocrinological and predisposition factors have also been considered, and familial tendencies to the disease have been noted.

False positive Wassermann tests have been found in some 20 percent of SLE patients, sometimes years before clinical symptoms of the disease appeared. So-called butterfly skin lesions, once thought to be the most typical characteristic of the disease, occur in less than 50 percent of the cases. Scaly skin eruptions of the face, ears, and scalp only are diagnosed as discoid lupus erythematosus; when the rash affects other parts of the body and when other tissues are involved, the disease is systemic.

SLE patients may suffer from various disorders, singly and in combination, and in no known consistent pattern of time or order. These include skin eruptions; anemia (80 percent); inflammation of joints, often indistinguishable from rheumatoid arthritis (90 percent); pleurisy, often resembling pneumonia and frequently complicated by infection (more than 50 percent); nervous system involvement, including fits, hallucinations, psychosis, and paranoia; acute gastrointestinal manifestations; enlarged spleen (15 to 20 percent) and lymph nodes; and a host of blood disorders.

This report is based on a statement from the National Foundation, New York, N.Y.

Diagnosis of SLE is often difficult because of the episodic manner in which the disease can appear over a period of years. A variety of symptoms are involved, and many symptoms mimic those of other diseases. SLE often resembles rheumatoid arthritis so closely that differential diagnosis is a problem. Finding the lupus erythematosus cell and making appropriate biopsies are helpful in confirming the diagnosis.

With an estimated 1,000 or more new victims of SLE being hospitalized each year in the United States, physicians are more and more concerned about treatment. There is no generally accepted treatment of choice. Some physicians favor regular high doses of steroids, such as cortisone, to reduce inflammation; others prefer low-steroid therapy combined with other measures.

Treatment of SLE is aimed at controlling and suppressing symptoms. Salicylates are used to relieve pain, particularly when joints are involved, and to reduce fever. Corticosteroids are used, sometimes in very high dosage, to reduce inflammation, but they have no effect on the basic cause of the lesions. Steroids often have dramatic results but may produce serious side reactions. Antimalarial drugs such as atabrine and chloroquine are used in treating SLE in its milder forms, especially arthritis and skin lesions, and also in treating patients on prolonged high-steroid regimens in whom it is desirable to reduce the steroid dosage. Appropriate antibiotics are used to treat complicating infections. Generally speaking, indicated standard measures are also used for various SLE manifestations. The most critical problems are encountered when renal involvement occurs. The apparent value of sustained high-steroid regimens in treating this condition needs confirmation.

Patients selected for the collaborative study must be willing to take part and must meet rigid criteria. Diagnosis of SLE with glomerulitis must be confirmed by kidney biopsy, and there must be no contraindications to high-dosage steroid therapy. For each patient information will be recorded on more than 300 variables. These will be reevaluated periodically, and the information will be recorded on punchcards for statistical analysis.

Half of the qualified patients, selected at random, will be kept on a regimen of 50 mg. or more of corticosteroids a day for 6 months. The other half will be treated with aspirin, antimalarial drugs, and moderate or low steroid doses, as needed, to control the symptoms and course of the disease.

Even though fewer than 100 patients are likely to be involved in this study during the first year, the collection and correlation of masses of data "could provide a goldmine of information," and Dr. Clark and the directors of the Arthritis Clinical Study Centers "are very hopeful that some vital clues to the SLE puzzle will turn up." They hope that results of the study "will take some of the guesswork . . . out of treatment problems and give us new guidelines for handling the disease."

The study will be conducted at Columbia University College of Physicians and Surgeons, Presbyterian Hospital, New York City; Johns Hopkins University, Baltimore, Md.; University of Texas Southwestern Medical School, Dallas; University of California School of Medicine, San Francisco; and University of Rochester School of Medicine, Rochester, N.Y.