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South Boston Scleroderma and Lupus Health Study

Questions & Answers

What is the South Boston Scleroderma and Lupus Health Study (SBSLHS)?

The South Boston Scleroderma and Lupus Health Study is a retrospective case-control study which sought to investigate the prevalence of these diseases in South Boston and their possible association with environmental and non-environmental factors.

Why was the SBSLHS conducted in South Boston?

In 1998, South Boston residents and then State Senator Stephen Lynch (now Congressman Lynch) contacted the Massachusetts Department of Public Health (MDPH) with concerns over a suspected increase in the number of individuals with systemic scleroderma in their community. The concerns focused on a perceived increase in the number of women who grew up in South Boston who had been diagnosed with scleroderma and the potential relationship with historical opportunities for environmental exposures in South Boston. Some community residents also expressed concern that the prevalence of lupus may also be higher than might be expected.

Why did MDPH evaluate both of these diseases in one study?

Clinical co-investigators advised MDPH that given the statistical challenges of conducting a study of scleroderma (a disease with relatively low prevalence) and the similarities in some factors associated with both scleroderma and lupus, combining the two diseases in one investigation might increase the chances of determining environmental contributions in a single geographic area.

Who conducted the study?

The MDPH's Bureau of Environmental Health (BEH) conducted the study in partnership with clinical researchers and rheumatologists from Boston Medical Center, Tufts Medical Center, and the South Boston

Community Health Center. In addition, MDPH established the South Boston Community Advisory Committee (CAC) to enhance outreach and provide ongoing communication about the various milestones of the study.

What is scleroderma?

Scleroderma is a chronic connective tissue disease. It is characterized by hardening of the skin and internal organs, mostly due to an over-production of collagen (the substance present in scar tissue). The word scleroderma literally means hard (“sclerosis”) skin (“derma”). Scleroderma mainly affects women of child-bearing years and early menopausal years (peak incidence is between 45 to 54 years of age).

How common is scleroderma in the general population?

Scleroderma is a rare disease, and no one knows exactly how many people have this disease. Based on studies conducted in the United States, in any given community at any given time, it is estimated that approximately 28 per 100,000 people may have scleroderma. The rate may be as high as 37 per 100,000 among white females and 43 per 100,000 among black females.

What is lupus?

Lupus is a chronic inflammatory disease. It is characterized by inflammation of various parts of the body, particularly the skin, joints, blood system and kidneys. Ninety percent of patients are women, and the usual age at onset is between 15 and 40 years. African American women are 3-4 times more likely to develop lupus than white women.

How common is lupus in the general population?

It has been estimated that anywhere from 14 to 150 per 100,000 people may have lupus in the United States. Recent estimates based on national population data have found approximately 54 diagnoses per 100,000 in the general population and 100 per 100,000 among females.

How was the study conducted?

Prior to the start of the study, a detailed scientific protocol was developed. MDPH/BEH researchers collaborated with physicians at Boston Medical Center and Tufts Medical Center to develop the protocol. The protocol was reviewed and approved by the MDPH Human Research Review Committee in late 1999.

How did MDPH identify current and former residents who had been diagnosed with scleroderma or lupus?

MDPH and the Community Advisory Committee, often helped by residents, conducted major outreach efforts beginning in 2000 to identify and encourage residents with these diseases to participate in the study. This included both current and former residents. Outreach efforts included contact with local hospitals and rheumatologists, fliers, community fundraising events, reporting on local cable television and national network television, and newspaper/magazine articles.

Who participated in the study?

Current and former residents of South Boston who were diagnosed with scleroderma or lupus between 1950 and 2000 were included in the study. These individuals were required to be at least 21 years of age at the time of the study and have resided in South Boston for at least one year. A comparison population included people who were randomly selected from the 2001 South Boston residents list (an annual census compiled by the City of

Boston) and were matched with cases based on gender and age. These individuals were required to live in South Boston for at least one year prior to the diagnosis date of the case to which he/she was matched.

What kind of information was collected during the interviews for the study?

Throughout 2004, each study participant was given a questionnaire by a trained interviewer. Interviewers collected detailed information on residential history, occupational history, personal and family medical history, reproductive history, and hobbies and recreational activities involving certain chemicals. The study also investigated possible exposures to a number of historical sources of environmental pollution in the South Boston area including the Coastal Oil site, a former power plant, and other hazardous waste sites.

How many people participated in the study?

Of the 45 individuals with a confirmed diagnosis of scleroderma or lupus, 41 agreed to participate in the study. Of the 830 individuals selected as potential controls, 219 agreed to participate in the study. Of these 219 individuals, 154 met study eligibility criteria and were matched to cases.

What was the overall participation rate for the study?

The overall participation rate for the study was 22% (195 study participants/875 eligible population).

How much did the study cost?

The study cost approximately \$1,750,000 in state funding. In fiscal years 2000 through 2004, line item directives of \$300,000 in state funds were provided to support the South Boston study. Since that time, the MDPH/BEH operating account has supported the study activities necessary to complete the investigation.

Was the study peer reviewed?

Yes. The South Boston Scleroderma and Lupus Health Study was peer-reviewed by two national experts in scleroderma and lupus research: Dr. Maureen Mayes who is a rheumatologist, a professor of medicine at the University of Texas-Houston Medical School and the Director of the Scleroderma Clinic; and Dr. Glinda Cooper, an epidemiologist with the National Center for Environmental Assessment at the U.S. Environmental Protection Agency. Dr. Cooper's research focuses primarily on rheumatic autoimmune diseases and environmental exposures. Peer review comments were provided to MDPH in the fall of 2008. Additional analyses were conducted and the report was revised to address those comments in 2009.

What were the major findings of the South Boston Scleroderma and Lupus Health Study?

- The South Boston Scleroderma and Lupus Health Study found that the *prevalence* of scleroderma (the total number of people living with scleroderma) was higher than expected in South Boston based upon previous estimates in the medical literature.
- The prevalence of lupus was within ranges reported in the medical literature.
- The *incidence rate* (the number of people diagnosed each year) for each disease was within ranges reported in the medical literature.
- A previous diagnosis of rheumatoid arthritis was associated with scleroderma and lupus among current/former South Boston residents.
- A family history of specific rheumatic diseases was associated with increased risk of both scleroderma and lupus.

- The study did not find any geographic or temporal (time) patterns among individuals living/having lived in South Boston and being diagnosed with either scleroderma or lupus.
- No associations suggesting potential environmental exposures specific to South Boston were identified.
- However, it is important to note that the low participation rate may have limited the ability to detect associations between the risk of developing scleroderma or lupus and other environmental factors.

Are there any recommendations based on the study findings?

Based on the findings of this study, no recommendations for additional study of South Boston are being suggested. However, research currently underway involving clinical registries may better evaluate how human genes are affected by interactions with the environment and characterize the role of environmental factors.

How can I learn more about scleroderma registries?

While there are no population-based scleroderma registries in the United States, there are some clinical researchers who are recruiting patients diagnosed with scleroderma. A national registry is funded by the National Institute of Health (NIH). It is part of a research study of the genetics of scleroderma to identify genes that might predispose people to get this disease. The registry began at the University of Texas Health Science Center at Houston and is now funded to include other medical/research institutions across the country. More information can be found at: <http://www.sclerodermaregistry.org>. Recently, Boston Medical Center agreed to serve as a site for recruitment for the NIH-funded national scleroderma registry. Registration is open to individuals across the U.S. with systemic scleroderma who meet certain eligibility criteria. For more information about the Boston Medical Center's participation in the Scleroderma Family Registry and DNA Repository, please contact the Scleroderma Center at 617 638-4310.

Where can I obtain a copy of the South Boston Scleroderma and Lupus Health Study?

The full report can be found at www.mass.gov/dph/environmental_health, under News & Updates, or by contacting the MDPH Bureau of Environmental Health at 617-624-5757.