ACR and EULAR release new classification criteria for IgG4-related disease

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The American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR) released the 2019 ACR/EULAR Classification Criteria for IgG4-Related Disease. It is the first criteria developed specifically for this recently recognized disease.

A draft of the criteria was presented during the 2018 ACR/ARP Annual Meeting in Chicago. Since that time, the criteria team performed a second validation study, which confirmed the high sensitivity and specificity that was found in the first validation study.

IgG4-Related Disease (IgG4-RD) is an immune-mediated disease that may affect different organ systems and often mimics other diseases like Sjögren's syndrome, pancreatic cancer, granulomatosis with polyangiitis (GPA), giant cell arteritis (GCA) and systemic lupus erythematosus (SLE or lupus). Only recognized in the last 10 to 15 years, IgG4-RD can cause fibro-inflammatory lesions in nearly any organ or multiple organs. Estimates suggest that IgG4-RD affects 180,000 people in the United States and many more worldwide.

"IgG4-RD is now recognized to be a worldwide condition that is seen not only by rheumatologists but also generalists and sub-specialists of nearly every kind," said John H. Stone, MD, MPH, professor of medicine at Harvard Medical School and director of the international panel of experts who developed the new criteria. "Clinical trials are now being developed in IgG4-RD and investigators need criteria on which to base patients' inclusion or exclusion for such trials and other types of

investigation."

Classification criteria allow researchers to accurately identify patients for inclusion in clinical, epidemiologic and basic investigations. The panel of experts who developed the new classification criteria included investigators from rheumatology and other specialties from five continents, reflecting the worldwide impact of this disease.

In the criteria, classifying patients with IgG4-RD is a three-step process that carefully assesses data from four domains, which must make sense in the context of IgG4-RD. The process includes synthesizing information from the patient's clinical presentation, blood test results or serology, radiological findings and the pathology data. Few other diseases require such careful synthesis of various information to get an accurate diagnosis, and at this time, there is no single diagnostic test for the disease.

The 2019 ACR/EULAR Classification Criteria for IgG4-Related Disease were validated in a large cohort of patients and demonstrated excellent test performances. Dr. Stone feels they should be a highly useful contribution to future investigations in this disease, and will ultimately help improve the lives of patients with IgG4-RD.

"IgG4-RD is a disease that tends to afflict middle-aged to elderly individuals and often affects and damages the pancreas severely, making glucocorticoids a suboptimal therapy for this condition," Dr. Stone says. "Clinical trials will help develop targeted therapies that spare toxicities from conventional treatments. Investigators need to have criteria like this to determine whether a patient should be classified as having IgG4-RD."

Dr. Stone is a professor of medicine for Harvard Medical School and the Edward A. Fox Chair in Medicine at Massachusetts General Hospital.

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