

Researchers Develop First Integrated Clinical Management Scheme for Managing Alpha-1 Antitrypsin Def

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DENVER — Alpha-1 antitrypsin deficiency increases the risk of chronic obstructive pulmonary disease (COPD), liver disease, and several other conditions, but is often unrecognized. National Jewish Health pulmonologist Robert Sandhaus, MD, PhD, and Edwin Silverman, MD, PhD, of Brigham and Women's Hospital, offer the first concise, integrated clinical management scheme for healthcare providers managing Alpha-1 patients in the June 25, 2009, issue of *The New England Journal of Medicine*.

"This paper should offer a blueprint to medical providers managing Alpha-1 patients," said Dr. Sandhaus, who is widely recognized as one of the foremost experts on the disease.

Alpha-1 Antitrypsin Deficiency is a genetic disease that results in abnormally low levels of the protein alpha-1 antitrypsin. Alpha-1 antitrypsin is produced in the liver. Its main function is to protect the lungs from an enzyme that digests damaged or aging cells and bacteria. Unchecked, that enzyme can damage lung tissue. Abnormal alpha-1 proteins can also become trapped in the liver and cause damage there.

Experts estimate that only about 10 percent of people with alpha-1 antitrypsin deficiency are diagnosed. That is why one of the authors' primary recommendations is to test serum protein levels and confirm any findings with genetic tests for patients suspected of alpha-1 antitrypsin deficiency. Drs. Sandhaus and Silverman conclude that close monitoring of patients for the development or progression of lung or liver disease is required. Consultation with lung and liver disease specialists is also recommended. The diagnosis of alpha-1 can also be a strong motivator for patients to quit smoking and an excellent opportunity for physicians to counsel their patients to do so. Alpha-1 augmentation therapy, to replace the missing protein, should be considered, according to Drs. Sandhaus and Silverman.

"We hope these guidelines will raise awareness of alpha-1 antitrypsin deficiency among physicians and lead to better diagnosis and management of the disease," said Dr. Sandhaus.

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