Alpha-1 to protect the lungs. if these proteins

Alpha-1 Antitrypsin DeficiencyMitchell ClarkLutheran School of NursingNR20411/27/2017 Medlineplus (2017) defined Alpha-1 antitrypsin deficiency asa hereditary disease passed down from each generation that increases the chancesof lung and liver disease. Alpha-1 antitrypsin is a protein that the liverproduces in order to protect the lungs.

If these proteins are not the correctshape, they will get trapped inside the liver cells and are not able to reachthe lungs in order to safeguard them which is how Alpha-1 antitrypsindeficiency occurs (MedlinePlus, 2017). Carterand Tiep (2008), explained thatAlpha-1 antitrypsin (AAT) deficiency and the deficient amount of circulatingAAT, increases the possibility of a serious disease that affects the lung andliver. Originally it was thought that only people of northern European ancestrywere plagued by this illness. However that was not the case because it isMU1 now known that people are affectedall over the world. Detecting this deficiency prior to the development ofsevere lung disease is very important due to the fact that if therapy isstarted early it might limit the deterioration of organ function. The patientswith this disease should immediately stop smoking and avoid smoke-filled surroundings. These patients should also be told to avoid lung irritants, have better diets, workout, use precautionary methods against the disease, and become well-informed aboutthe condition and the signs and symptoms of exacerbations.

(Carter& Tiep, 2008)Medline Plus (2017), states that the signs of Alpha-1antitrypsin deficiency include shortness of breath, wheezingm2 , repeated lung infections, tiredness, rapid heartbeat upon standing, vision problems,

and weight loss. However, somepeople do not develop symptoms or any complications at all. In order to diagnose this disease aseries of blood tests and genetic tests are used for diagnosis. The treatmentsfor this disease include medicine, pulmonary rehab, extra oxygen, and if severeenough a lung transplant. The best approach to prevent or delay lung symptomsis to stop smoking (Alpha-1 Antitrypsin Deficiency, 2017). According toStoller (2017), Alpha-1 Antitrypsin Deficiency is controlled through genetictesting and counseling. Once someone is diagnosed with this disease it isimportant to have the rest of the family tested.

This disease is inherited, andfamily members of these patients areat a higher risk for having Alpha-1 antitrypsin deficiency and for developingassociated diseases. These individuals should be counseled onsmoking cessation and careers that involve less dust exposure. Tanash, Nilsson, Nilsson, andPiitulainen (2010), published a study where 315 people with AATwere studied and 24% of them died. The smokers in this study had a higher riskof death than those who have never smoked, and among the cases that werescreened, the rate of those that died was significantly higher in smokers thanin the general population. According to Anariba’s (2017), articlealpha1-antitrypsin deficiencies earliest definition was by Laurell and Erikssonin 1963. Laurell noticed that there was not a band of alpha1-protein in five of the fifteen hundred serum proteins that were given to his laboratoryin Sweden. Both scientist then found that three of the five patients that hadthe band missing on the protein had emphysema when they were younger, and someeven had a family members who also had emphysema.

Thus the symptoms of alpha-1 antitrypsin deficiencywere recognized and they included: that there was no protein in the alpha-1 section of the SPEP, emphysema thatpresents itself at an early age, and a family history of this disease. Konvalinka(2010), debated the implications for having Alpha-1 Deficiency and thedifficulties associated with it. This article is about the author's work withpanniculitis, which is an infection of the layer of fatty and fibrous tissue layersof our epidermis that people with the AAT deficiency sometimes get. Accordingto Konvalinka (2010), the inflammation is located beneath the skin in ahoneycomb shape. She declared that the honeycomb pattern of fat below the skin mightbe produced by the absence of the Alpha-1 protein. Konvalinka (2010), startedher study by giving a brief overview of the deficiency. She goes on to argue thatpanniculitis associated Alpha-1 deficiency differs from typical panniculitis. She stated that the AATD Panniculitis usuallyoccurs in young adults and starts with painful nodules usually on the thigh orthe buttocks, but can occur anywhere on the body, which are warm, red, andtender.

(Konvalinka, 2010, p. 24) Theresearch findings of Konvalinka (2010), could be crucial in a clinical setting dueto the fact that Alpha-1 Deficiency could present with a similar diagnosis as somebodywho does not have the deficiency but may have other indicators of a disease. Thedifferences among the two types of panniculitis are vital for those in themedical field to differentiate. Not only in the case of panniculitis, but inorder to recognize that people with the AAT deficiency may show different signsand symptoms than those without the absence of this protein.

Early recognitionand diagnosis of this disease is of the up most importance. Konvalinka (2010), went on to discuss new improvements in accessibility of testing for the deficiencyand some of the available treatments for it, such as plasma exchange and antibioticsto control the panniculitis. The research article concluded by pointing outthat difficulties related to the alpha 1 deficiency, like panniculitis, aresometimes the only way to be tested for a protein deficiency.

This is a greatway for those working in the medical field to better understand those livingwith the deficiency. Doctors and nurses need to be more aware of this proteindeficiency, due to the fact that patients in their care could be carriers or beaffected with the disease.
Apersonal interview with Amy Wallen, a nurse practitioner who used to work atWashington University in the renal division, was an excellent source of informationon the subject.

She said that, “ This disease stood out to me more than othersbecause it took a patient who was otherwise young and healthy and took them tothe point of a debilitating illness.” (Personal communication, November 20th, 2017) Amy now works in a correctional facility in Farmington taking care ofpatients. Though she has only had a few patients in the prison with this diseaseMU3 , the treatment is no different. EMU4 venthough it is difficult sometimes to arrange the care they need due to safetyand security reasonsMU5 , there has never been a time when a prisoner in need was not given the help heneeded.

When asked what treatment she believes is the most effective shestates, “ I think the most effective way to treat the disease is to preventcomplications by treating problems early, avoiding alcohol, smoke andpollutants and make yourself knowledgeable of the common problems that canoccur.” It is important to have knowledgeable medical professionals like Amy inorder to take care of these patients in the best way possible.

Alpha-1Foundation (2017), stated one of the best ways that patients can cope with thisdisease is to join a support group and to talk with people who are going throughsome of the same problems. One of the main ways patients get into contact witha support group is through this foundation. This foundation not only helpspeople get into contact with a support group, but also promotes research andthe advancement of new treatments and the improvement of the quality of lifefor Alpha-1 patients.

This organization works with the National Institutes ofHealth, the Food and Drug Administration, the people plagued byAlpha-1, and the pharmaceutical industry to speed up research andimprovement of therapies for this disease (Alpha-1 FoMU6 undation, 2017). Organizations like this are a great help to society, not only do they helpthose in need but also further scientific research while doing so. In conclusion Alpha-1 antitrypsinwas never a preventable illness but instead a hereditary illness that waspresent from birth to death. If theseproteins are decreased complications arise such as COPD, emphysema, pancreatitis and liver disease.

There is hope for the people affected with thedisease though. Organizations like the Alpha-1 Foundation and knowledgeablemedical staff such as Amy Wallen impact and benefit the population. They bothhope to serve by gaining

more knowledge and constantly seeking new treatmentsand options for those living with the deficiency. Even though through treatmentand counseling this disease is manageable it is still a heavy burden to placeon a family. Patients rely on their families for support and guidance. It isimportant for medical professionals to be competent on the disease process. Coping with Alpha-1 Antitrypsin Deficiency can be difficult, but throughperseverance and treatment this disease will not define the patient’s life.