Alpha 1 Antitrypsin MZ Information & Research

News & Research Update

Jun 22, 2024

Dear Subscribers,

The Last three newsletters we covered three different therapeutic medication in development:

RNAi and RNAe (messenger) therapeutics which is not a permanent change and needs regular injections.

> RNAi which inhibits the expression of the "Z" mutation of the SERPINA1 gene and as such reduces the misfolded "Z" AAT protein production in the liver, and provides a solution for the liver issues we encounter (including the second order issues as a result of it).
>
> RNA editing, which allows the liver to make more of the correct "M" protein instead of the misfolded "Z" protein, and is as such a solution for both liver, lungs, your immune system and anti-inflammatory issues.

DNA editing therapeutics which is a permanent change in your DNA, and corrects the Z mutation and turns it back into the normal "M", which has more or less the same results as RNA editing, but is permanent and does not require regular injections.

There are however developments to increase the Alpha 1 Antitrypsin levels without RNA and DNA Editing. These are not only under development for Alpha 1 patients, but also for patients with e.g. auto immune issues. Below an abstract of the other issues around an Alpha1 Antitrypsin deficiency and the involvement of A1AT in other Alpha1 morbidities.

Alpha-1 antitrypsin in autoimmune diseases: Roles and therapeutic prospects

Source; https://pubmed.ncbi.nlm.nih.gov/35803133/

Alpha-1 antitrypsin (A1AT) is a protease inhibitor in serum. Its primary function is to inhibit the activity of a series of proteases, including proteinase 3, neutrophil elastase, metalloproteases, and cysteine-aspartate proteases.

In addition, A1AT also has **anti-inflammatory**, **anti-apoptotic**, **anti-oxidative stress**, **anti-viral**, and **anti-bacterial** activities and plays essential roles in the regulation of **tissue repair** and **lymphocyte differentiation and activation**.

The overactivation of the immune system characterizes the pathogenesis of **autoimmune diseases**. A1AT treatment shows beneficial effects on patients and animal models with autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus.

As you can see in this article, your Alpha1 Antitrypsin deficiency is not only affecting your lungs and liver, & gastric tract, but also has a negative effect on your immune system.

One example is your Lymphocytes (T cells), which are important to fight off viruses in your body. You may have noticed that you were/are more sick compared to others, and that it takes more time to fight a virus infection. So, your defense system is impaired, and it also takes longer, and is more severe, because you have a lower anti-inflammatory level. (Note: I am a good example because my Lymphocytes are far below the minimum…)

Please note that your Lymphocytes are typically not tested as part of a regular blood test, were they only test the number of Leukocytes (white blood cells) but not the composition of the Leukocytes like the lymphocytes etc. So, something to be aware off, and may be ask for.

In previous issues we already mentioned the involvement of A1AT in tissue repair and the added risk for a potentially deadly aneurism when you are getting older, so you may want to check this on a regular basis.

In conclusion, when we look at the complete picture of an Alpha 1 Antitrypsin deficiency you need to consider all morbidities and not only lung and liver. Please note that the severity may vary from person to person and that age plays an important role.

And like always, enjoy the ride !!

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