Treatment of Cystic Fibrosis with Penicillamine. A Hypothesis Based on the Physiology of Mucus and Sweat

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This work describes the possibility of treating cystic fibrosis with penicillamine, it would block the copper and other trace metals involved in the formation of mucus and sweat, and could decrease or improve their formation.

Cystic fibrosis, also called mucoviscidosis, is an inherited disease characterized by increased production of mucus in the lungs and digestive system and production of altered sweat.

It has been shown that in the mucus (sputum) of patients with cystic fibrosis, there are present trace metals, mostly iron and copper [1], and also zinc [1].

Interestingly, patients with cystic fibrosis have decreased their serum copper, which does not improve significantly after administration of this metal orally [2], this may be because the copper in these patients is been used for the formation of mucus [1]; sweat is also altered in patients with cystic fibrosis, maybe because the same mechanism: utilization of copper for its formation. There is enough evidence to demonstrate that trace elements, mainly copper and zinc, are present in sweat of normal people [3], so they must be present also in sweat of patients with cystic fibrosis in any stage of the disease. It is also important to remark that patients with Wilson’s disease produce significantly smaller volumes of sweat [4], apparently, copper also participates in the physiology of sweat glands, the same glands affected in cystic fibrosis.

For all this evidence, the possibility of treating cystic fibrosis with penicillamine could succeed, as is in Wilson disease; penicillamine is one of the most effective chelators [5], and it would block the copper [5] and other trace metals involved in the formation of mucus and sweat, including zinc [6] and doing this, patients with cystic fibrosis would improve producing less mucus and sweat, or, better quality of both of them.

Penicillamine is effective as a treatment for rheumatoid arthritis [7] and has been used in patients with cystic fibrosis for the treatment of related arthritis [8], but so far, penicillamine has not been used to decrease mucus and sweat production by blocking the copper hypothetically involved in the production of them.

References

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