Website address:
https://www.gesundheitsindustrie-bw.de/en/article/press-release/restarting-the-muscular-engine-research-prize-awarded-to-two-medical-practitioners-from-ulm-and-heid

Restarting the muscular engine – Research Prize awarded to two medical practitioners from Ulm and Heidelberg

Dr. Karin Jurkat-Rott, associate professor at the Institute of Applied Physiology at the University of Ulm, and Dr. Marc-André Weber, associate professor and chief physician in the Department of Radiology at the Heidelberg University Hospital, were awarded the Eva Luise Köhler Research Prize for Rare Diseases in Berlin on 1st March 2010. The prize was awarded by Luise Köhler in the presence the German Federal President Horst Köhler, the German Federal Minister of Health and the Spanish Crown Princess Letizia of Asturias. The prize was awarded to an innovative strategy for the treatment of a rare muscular disease characterised by periodic paralysis.

The medical practitioners received the prize of 50,000 euros from the Eva Luise and Horst Köhler Foundation (est. March 2006) for their innovative approach towards the improvement of the medical therapy of patients suffering from hypokalemic periodic paralysis (HypoPP). This rare muscular disease is one of over 6,000 rare diseases affecting around four million people in Germany. It is difficult to treat rare diseases because the handful of drugs available that might help are not yet approved for human application due to the time-consuming approval procedures for special indications.

Drastic changes of life

"Although muscular diseases are quite rare, progressive muscle atrophy nevertheless drastically affects the life of sufferers who become more and more immobile," said the prizewinner from Ulm. Dr. Karin Jurkat-Rott went on to add that muscular diseases also have a huge social and economic impact on relatives of the sufferer and on society as a whole. "There are numerous causes of such diseases, ranging from inflammation to hereditary factors."

There are currently no therapeutic means available that might be able to halt the process of progressive muscle atrophy. "This is also the case for the group of hereditary diseases that are caused by defective ion channels in the cell membrane," said the two medical practitioners, who have worked together in this field of research for many years.

Like a battery that is discharging

Dr. Marc-André Weber, chief physician in the Department of Diagnostic and Interventional Radiology at the University Hospital of Heidelberg, and Dr. Karin Jurkat-Rott, University Hospital of Ulm (on the right), are awarded the Eva Luise Köhler Research Prize for Rare Diseases by Luise Köhler, the Spanish Crown Princess Letizia of Asturias and the German Federal President Horst Köhler.

The two scientists describe their work using an analogy. They see the diseased muscle as a battery that is leaking and discharging. This battery is therefore unable to drive the muscle engine and this results in paralysis, rendering the patients immobile. One example of muscular diseases of this kind is hypokalemic periodic paralysis: HypoPP patients suffer periodically from paralysis, which typically occurs in the early hours of the morning after having consumed a rich meal the previous evening. Sufferers are unable to move their arms and legs and they cannot get out of bed. In the best case, this only lasts for a few minutes or hours. However, as sufferers get older, permanent muscle weakness requiring the use of wheelchairs can develop.

Paralysis caused by a decrease in the potassium level

A decrease in blood potassium levels is the major cause of periodical bouts of paralysis. Once the potassium levels return to normal again, young patients regain their mobility. A normal potassium level therefore counteracts the leaking of the “muscle battery”, which explains why young patients with a normal potassium level have normal strength levels.

"The disease symptoms are often misjudged," explained Dr. Jurkat-Rott adding that many patients tend to see a psychiatrist rather than a neurologist. The disease has an estimated prevalence of 1:100,000. However, due to the aforementioned reasons, it is assumed that there is quite a large number of unknown cases.

Age favours muscle atrophy

With increasing age, about 50% of all patients suffering from muscle atrophy begin to find it difficult to walk, and maybe even become dependent on wheelchairs. Magnetic resonance imaging is able to detect the accumulation of sodium and water in affected patients. This is similar to the accumulation of water, i.e. the thinning of sulphuric acid in an old car battery. Once a critical value has been reached, the car engine can no longer be started. Thus, both the cold and age are unfavourable factors for leaking "muscle batteries" and old car batteries.

Surprise: muscular strength can be restored

It has previously been assumed that muscle atrophy, which gets worse as people get older, cannot be stopped or restored. Therefore, the scientists were surprised to see that getting rid of water and sodium from the muscle cells of HypoPP patients leads to the restoration of muscle strength. This is made possible through the use of some of the known diuretic drugs, and patients can then follow training programmes to build up their muscles. Dr. Weber and Dr. Jurkat-Rott used an acetazolamide therapy to treat two wheelchair-bound young women and this resulted in them being able to walk again.

As part of their project, the scientists have plans to test further drugs with fewer side effects with the aim of preventing the degeneration of muscles of HypoPP patients in the long term.

Direct measurement of muscle strength

The scientists will use the prize money to measure for the first time ever the recharging of the muscle battery in living people. This requires them to determine the distribution of chloride in the blood and the muscle cells. In order to achieve this, the researchers are planning to use a highly modern magnetic resonance imager (MRI) equipped with a special measurement technology (a 35chlorine coil). This coil will have to be manufactured specifically for this application.
The imager will help the scientists to determine hydrogen and sodium signals as well as the signal produced by chlorine in the muscle cells. The concentrations of these elements in the blood can be measured easily in routine laboratories.

"This system will allow us to assess the success of treatments involving different diuretic drugs after a very short period of therapy," said Dr. Weber. Besides the diuretic drug that the researchers have already tested there are substances that are able to additionally elevate the potassium levels in patients suffering from hypokalemic periodic paralysis. These drugs, of which some are already approved for human application, will potentially have a dual effect.

Eva Luise Köhler Research Prize for Rare Diseases

The Eva Luise and Horst Köhler Foundation has been awarding the 50,000-euro Research Prize for Rare Diseases since 2008 in cooperation with the German Alliance of Rare Diseases (ACHSE). The objective of the prize money is to boost and promote research into rare diseases in Germany in order to improve the diagnosis, prevention, medication and therapy of affected people. Rare diseases is an issue close to the heart of the Spanish Crown Princess Letizia, which is why it was natural for her to take part in the prize award ceremony in Berlin. The Foundation selected Dr. Marc-André Weber and Dr. Karin Jurkat-Rott's research from a total of 61 applications.

Literature:

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