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Mightier mouse offers hope to muscular dystrophy sufferers

Ian Sample, science correspondent

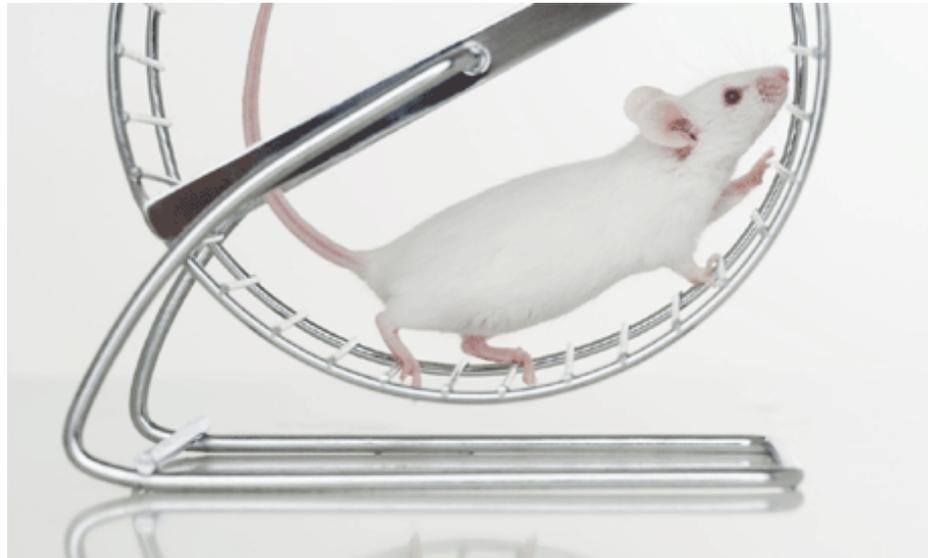
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Ten years after creating the first real life Mighty Mouse, scientists have unveiled an even mightier version.

The research has shed more light on how muscles develop in animals, and is expected to lead to new treatments for serious wasting diseases such as muscular dystrophy, and larger, leaner cattle for farmers.

The mice appear to be healthy but are substantially larger and weigh up to twice as much as normal mice, the researchers said.

Scientists at Johns Hopkins University in Baltimore, Maryland, revealed their first beefed-up mice in 1997. They were bred without a gene that produces a protein called myostatin, which limits muscle growth. As the mice grew they put on twice as much muscle mass as expected, dwarfing other mice in their cages.

In the latest research the team found that boosting the levels of another

protein, follistatin, doubled the muscle-making effect in mice which already lacked myostatin. "If you look at them they seem completely normal apart from their size. They're huge," said Se-Jin Lee, lead scientist on the project.

Inspections of the supermice revealed that their muscles were typically four times more bulky than normal. Individual muscle fibres were 117% larger, and they had 73% more fibres, according to details in the journal PLoS One.

"There's a lot of focus on using these findings to treat muscle wasting diseases, and also muscle loss associated with ageing," said Professor Lee. "It might also be possible to boost meat yields in cattle which haven't already been bred to optimise their muscle mass."

Some breeds of cattle and sheep naturally lack a myostatin gene and have been bred to produce lean animals.

One drug that blocks myostatin is already in clinical trials for the treatment of muscular dystrophy, while another is capable of boosting follistatin levels.

There are about 60 types of muscular dystrophy and related neuromuscular conditions that cause a steady deterioration of muscles and nerves. The conditions are untreatable, largely inherited, and affect more than 150,000 people in Britain, according to the Muscular Dystrophy Campaign.

Scientists believe follistatin blocks proteins in the body that prevent an animal becoming musclebound. "These animals are muscly but very lean. For humans, and lab animals, it's not much of a problem because we have ready access to food, but in the wild being too muscly puts a demand on the amount of calories you need to take in, so a balance is needed," Prof Lee said. "These findings show that the capacity for increasing muscle growth by targeting these pathways is much more extensive than we have appreciated."