

Researchers find link to failing heart in muscular dystrophy

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(Medical Xpress)—In a world first, researchers at The University of Western Australia have discovered a communication breakdown may be responsible for causing heart failure in people with muscular dystrophy.

According to a study, mice that lacked the protein dystrophin had impaired functional communication between a [calcium channel](#) in the muscle cell membrane and the [mitochondria](#) that are responsible for producing the energy.

It's understood that the hearts of muscular dystrophy patients fail but the mechanisms are unknown. It has also been previously reported that a lack of the protein dystrophin (responsible for muscular dystrophy) alters calcium channel function. In the latest study the researchers have identified a link between the channel and the mitochondria that involves dystrophin. In muscular dystrophy the functional communication between the channel and the mitochondria is impaired.

"We had previously shown that the L-type calcium channel could alter [metabolic activity](#) in the heart," Lead author Professor Livia Hool, explained. "The L-type calcium channel is widely recognised as being important for regulating the heart beat and contraction. We had demonstrated an additional role for the channel in regulating metabolic activity. This involved altered calcium influx but part of the response also involved proteins that maintain cell structure. This study demonstrated that when dystrophin is absent and the cell structure is in disarray, the communication between the channel and the mitochondria becomes disrupted leading to heart failure."

"This is an important finding, because it provides insight into a mechanism for the development of heart failure in muscular dystrophy," Professor Hool said. "In addition, we have demonstrated that treatment of the mice with morpholino oligomer peptides that induce functional expression of

dystrophin rescued metabolic activity in the mice.

"This study is exciting because it validates our hypothesis that the channel maintains normal metabolic activity by communicating with the mitochondria through the structural proteins. In [muscular dystrophy](#) hearts the communication is impaired because dystrophin is lacking.

"Our study also has significant implications with respect to understanding mechanisms of other types of heart failure. We are currently characterising metabolic activity in a number of animal models of human [heart failure](#) using the same approach."

The ground-breaking research is funded by the National Health and Medical Research Council of Australia, Australian Research Council of Australia, Heart Foundation of Australia, National Institutes of Health USA and Muscular Dystrophy Association USA. The collaborative group also holds NHMRC Project Grants to further improve uptake of the oligomer peptides into the heart so that therapy can be optimised in boys with Duchenne Muscular Dystrophy.

The study was published in the *Proceedings of National Academy of Sciences* .

Provided by University of Western Australia

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