



## Role of Sphingolipids in Muscular Dystrophy Disease

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### DESCRIPTION

Muscular dystrophy is a screen term for conditions where gene mutations affect in progressive weakness and breakdown of skeletal muscles. About half of all muscular dystrophy cases involve Duchenne muscular dystrophy (DMD). DMD arises from a mutation of the gene that codes for dystrophin, a protein supports muscle structure by anchoring the cytoskeleton of muscle cells with their cytoplasm, the sarcolemma. Mutations of dystrophin affect various natural pathways causing the hallmark symptoms of Duchenne muscular dystrophy compromised cells membrane integrity, aberrant calcium homeostasis, chronic inflammation, fibrosis, and disabled tissue alteration.

Discovered in 1870 and named after the noted Sphinx, sphingolipids are a group of bioactive lipids allowed to be involved in cell signaling, and, suddenly, numerous of the symptoms present in DMD. Thus, the investigators asked whether the synthesis of sphingolipids can be altered in DMD- and if so, if they can be causally involved in the pathogenesis of DMD. Sphingolipids are found in creatures, plants, fungi, and some prokaryotic organisms and viruses. It's a group of bioactive lipids thought to be involved in cell signaling, and, unexpectedly, numerous of the symptoms present in Duchenne muscular dystrophy (DMD).

First, they begin those mice with DMD show an accumulation of intermediates of sphingolipid biosynthesis. This was formerly an indication that sphingolipid metabolism is abnormally increased in the environment of muscular dystrophy. Next, the experimenters used the compound myriocin to block one of the crucial enzymes of the sphingolipid *de novo* synthesis pathway. Blocking synthesis of sphingolipids canceled the DMD-affiliated loss of muscle function in the mice. Digging deeper, the

experimenters plant that myriocin stabilized the development of muscular calcium, and reversed fibrosis in the diaphragm and heart muscle. At the same time, blocking the conflation of sphingolipids also reduced DMD- related inflammation in the muscles by moving the vulnerable macrophage cells off their pro-inflammatory state and pushing them towards an anti-inflammatory bone.

The experimenters wanted to know whether pharmacological inhibition of the sphingolipid conflation pathway could also restore muscle function and meliorate symptoms of DMD in the mouse model. They used the emulsion myriocin to block one of the crucial enzymes of the sphingolipid *de novo* conflation pathway in mdx mice. Myriocin is an asset of serine palmitoyltransferase (SPT), the first and rate-limiting enzyme of the sphingolipid biosynthesis pathway. Creatures were given intraperitoneal injections of the medicine three times a week, for six months.

The results verified that inhibiting synthesis of sphingolipids canceled the DMD-affiliated loss of muscle function in the treated mice. Digging deeper, the experimenters also found that myriocin stabilized the development of muscular calcium, and reversed fibrosis in the diaphragm and in heart muscle. Myriocin, an inhibitor of SPT, strongly reduced the abundance of sphingolipid intermediates and reversed multiple DMD-associated, fundamental pathogenetic pathways, including aberrant  $Ca^{2+}$  homeostasis, compromised sarcolemmal membrane integrity, satellite cell imbalance, habitual inflammation, and fibrosis. The observation that by inhibiting the sphingolipid *de novo* synthesis pathway, we were suitable to reverse not only one but several pathophysiological pathways involved in DMD points to the high remedial eventuality of targeting sphingolipid synthesis pathway.

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